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Editorial: CORONAVIRUS AND THE OTORHINOLARYNGOLOGIST

The world finds itself in a unique situation at this moment. The 2019 Novel Coronavirus pandemic seems to have pushed the globe into an unprecedented crisis. COVID-2019 was a pandemic which began from Wuhan in China. The World Health Organization declared it a public health emergency on January 20, 2020. As of 27 March 2020, over 24000 deaths have occurred the world over and the numbers seem to be rising exponentially by the day.

As Otorhinolaryngologists, we find ourselves working closely with the patients, as the rapid spread of the disease continues. It is common practice for patients presenting with upper respiratory tract symptoms like those seen in COVID-19, to see an otolaryngologist as their primary care physician. Indeed, the first physician fatality which was documented globally as a consequence of COVID-19, was that of an otorhinolaryngologist in Wuhan on 25 January. It is thus paramount that orolaryngologists globally stay abreast of this outbreak and practice essential precautionary measures, which are crucial in protecting both themselves as well as their patients.

As the crisis seems to worsen, physicians all over the world have been forced to reluctantly reduce their services with regard to elective surgeries. Although essential emergency and oncologic surgeries are still proceeding, but the gravity of the current situation is causing major inconvenience to the patients and their relatives who often rush to hospitals only to be told that admitting patients has been put on hold to make space for prospective COVID-19 patients.

One particular area of concern in our field are aerosol-generating procedures, including tracheostomies and fiberoptic laryngoscopies. There seems to be no clear directive as to what precautions need to be taken in this regard. As for now, those patients clinically triaged as being non urgent are being exempt from undergoing any active procedures.

In many settings, otolaryngologists are being called upon to perform nasopharyngeal and oropharyngeal swabs to aid in the diagnosis of coronavirus suspects. In the Indian Government Hospital framework, even good quality Personal Protective Equipment (PPE) and appropriate masks are in short supply. Young doctors, being asked to look after coronavirus patients in the absence of adequate PPE, cannot help but feel that they are 'martyrs in waiting'.

It is difficult to fathom how a sub microscopic infectious agent has brought the entire world to a standstill. These are the worst of times, indeed. Yet, there is hope. Frantic research is going on globally to come up with a vaccine to tackle COVID-19. Hydroxychloroquine has shown some promise as a prophylactic agent. Ours being a tropical country, stands to benefit from the onset of warmer weather which may curb the spread of the virus. Eventually, our best bet against this contagious virus may be the development of herd immunity. Until then, it is imperative that we, as both doctors and citizens, play our part in combatting this deadly virus.

INNOVATIVE ALTERNATIVES OF PPE IN COVID-19 PANDEMIC - OUR EXPERIENCE

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Introduction & Background:

In the current scenario of rapidly spreading pandemic infection due to the coronavirus, SARS-CoV2, otolaryngologists and surrounding staff are especially vulnerable to viral transmission directly through mucus, blood, and aerosolized particles when examining or operating. There is evolving evidence from China, Italy, and Iran that otolaryngologists are in the group with the highest risk of contracting the virus while performing upper airway procedures and examinations, if not using appropriate Personal Protective Equipment (PPE).

Until the supply of testing materials catches up with the accelerating demand for testing, there will be circumstances that the COVID-19 status of these patients is unknown.

Therefore, when a detailed examination or surgical procedure is necessary for urgent or emergency care and the -19 status of the patient cannot be confirmed, then the patient should be handled as if they are COVID-19 positive. This consideration should apply regardless of whether one is in an office, hospital, or operating room setting. Furthermore, the provider and surrounding staff must have the necessary PPE.

As the coronavirus disease 2019 (COVID-19) pandemic accelerates, global health care systems have become overwhelmed with potentially infectious patients seeking testing and care.

Preventing spread of infection to and from health care workers (HCWs) and patients relies on effective use of personal protective equipment (PPE)—gloves, face masks, air-purifying respirators, goggles, face shields and gowns. A critical shortage of all of these is projected and has already developed in Indian hospitals.

PPE, formerly ubiquitous and disposable in the hospital environment, is now a scarce and precious commodity in many locations when it is needed most to care for highly infectious patients. An increase in PPE supply in response to this new demand will require a large increase in PPE manufacturing, a process that will take time many health care systems do not have, given the rapid increase in ill COVID-19 patients.

It's a challenging problem, and we're working to solve it in several ways. When PPE supplies cannot be filled by a health care facility's normal suppliers and the state cannot find a vendor, the best option left to doctors across the globe is to innovate and come up with temporary alternative solutions to PPE using locally available materials, at least till the time proper and convention PPE is made readily available to them. This article discusses various such alternatives being used at our hospital & some other such innovations from across the globe.

MATERIALS & METHODS:

The team at Department of ENT, Calcutta National Medical College & Hospital, Kolkata during the month of March 2020, focused on innovating & researching about temporary alternatives to personal protective equipment (PPE), in response to the current trend of PPE scarcity around the world.

This article discusses various such alternatives used at our hospital & other such innovations from across the globe.

The team designed two types of easy to make head gear (fig.2&3), which can be used while doing aerosol generating procedures like emergency tracheostomy, sample collection etc.

A group of nurses were specially allotted for preparation, sterilization & proper disposal of the head gears in advance. Sterilisation before the usage was done by either ethyl alcohol (92%-95%) or 1% hypochlorite solution. After usage it was disposed of in yellow cloured plastic bags /containers.

Materials Used:

A) For Model "A" face shield head gear:

- 1) OPG Plastic Sheets
- 2) Thick ribbon thread (30 cm)
- 3) Scissors
- 4) Cellophane tape

B) For Model "B" head gear:

- 1) Transparent plastic packet(50 cm(L) x 60 cm(B))
- 2) Surgical Mask
- 3) Scissors
- 4) Cellophane tape.

DISCUSSION :

In its current guidance to optimize use of face masks during the pandemic, the Center for Disease Control and Prevention (CDC) identifies 3 levels of operational status: conventional, contingency, and crisis¹ During normal times, face masks are used in conventional ways to protect HCWs from splashes and sprays.

When health care systems become stressed and enter the contingency mode, CDC recommends conserving resources by selectively cancelling non-emergency procedures, deferring non-urgent outpatient encounters that might require face masks, removing face masks from public areas, and using face masks for extended periods, wherever feasible.

When health systems enter crisis mode, the CDC recommends cancellation of all elective and non-urgent procedures and outpatient appointments for which face masks are typically used, use of face masks beyond the manufacturer-designated shelf life during patient care activities, limited reuse, and prioritization of use for activities or procedures in which splashes, sprays, or aerosolization are likely².

When face masks are altogether unavailable, the CDC recommends using of face shields without masks, taking clinicians at high risk for COVID-19 complications out of clinical service, staffing services with convalescent HCWs presumably immune to SARS-CoV-2 (severe acute respiratory syndrome coronavirus 2), and use of homemade masks, perhaps from bandanas or scarves if necessary. Most of the countries globally are rapidly entering crisis mode.

WHO³ says current global stockpile is insufficient, particularly for medical masks masks and respirators;

the supply of gowns and goggles is soon expected to become insufficient as well. Surging global demand – driven not only by the increasing number of COVID-19 cases but also by misinformation, panic buying, and stockpiling – will result in further shortage of PPE globally.

The American College of Surgeons (ACS) released a Statement on PPE Shortage during the COVID-19 Pandemic⁴ in which they recognized that many healthcare facilities are facing shortage of personal protective equipment (PPE), including face masks, gowns, and respirators. The ACS believes it is essential that surgeons, nurses, anesthesiologists, and other health care personnel be able to speak freely, without fear of retribution, as they seek to find a solution to accessing PPE. We advise surgeons to speak with their institutional leadership on these matters and to be able to direct any concerns to the ACS, with the goal of protecting themselves and their colleagues.

Suggested minimal PPE set :

The suggested minimal PPE set protects from contact, droplet and airborne transmission⁵ The composition of the set is described in Table 1.

Table 1: Composition of Minimal PPE set

	Protection	Suggested PPE
1	Respiratory protection	FFP2 or FFP3 respirator (valved or non valved version)*
2	Eye protection	Goggles (or face shield)
3	Body protection	Long-sleeved water-resistant gown
4	Hand protection	Gloves

*(In case of shortage of respirators, the use of face masks (surgical or procedural masks) is recommended. When this type of PPE is used, the limitations and risks connected to its use should be assessed on a case-by-case basis.)

Respiratory protection:

The respirator protects from the inhalation of droplets and particles. Because different types of respirators fit differently between users, the respirator requires a fitting test.

The use of class 2 or 3 filtering face-piece (FFP) respirators - FFP 2 or FFP 3, when assessing a suspected or

managing a confirmed case. An FFP3 respirator should always be used when performing aerosol-generating procedures.

Face masks (surgical masks) mainly protect from exhaled droplets⁵; their use is recommended in case of shortage of respirators and on a case-by-case assessment. Surgical masks do not require fit testing.

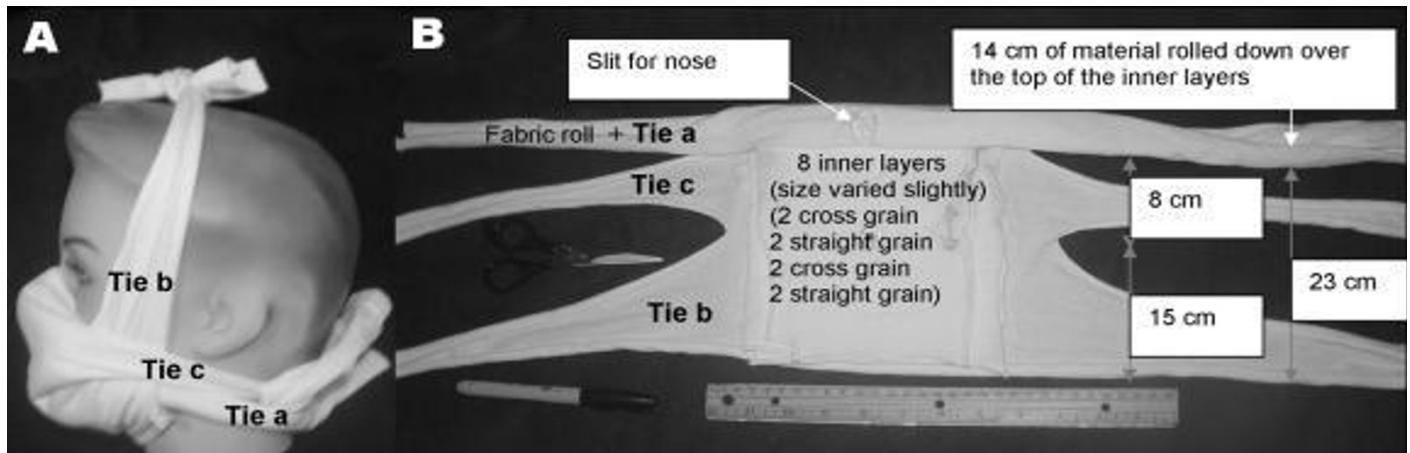


Figure 1 : Prototype mask. A) Side view, B) Face side. This mask consisted of 1 outer layer ($\approx 37 \text{ cm} \times 72 \text{ cm}$) rolled and cut as in panel B with 8 inner layers ($< 18 \text{ cm}^2$) placed inside (against the face).

Virginia M in a letter(6) described a simple, locally made, washable mask, which may be a solution if commercial masks are not available. A Hanes Heavy-weight 100% preshrunk cotton T-shirt (made in Honduras) was boiled for 10 minutes and air-dried to maximize shrinkage and sterilize the material in a manner available in developing countries. A scissor, marker, and ruler were used to cut out 1 outer layer ($\approx 37 \times 72 \text{ cm}$) and 8 inner layers. The mask was assembled and fitted as shown in Figure 1 . A fit factor is the number generated during quantitative fit testing by simulating workplace activities (a series of exercises, each 1 minute in duration).

The Portacount Plus Respirator Fit Tester with N95-Companion used for the test is an ambient aerosol instrument that measures aerosol concentration outside and inside the prototype mask. The challenge agent used is the ambient microscopic dust and other aerosols that are present in the air. A commercially available N95 respirator requires a fit factor of 100 to be considered adequate in the workplace. The prototype mask achieved a fit factor of 67 for 1 author with a Los Alamos National Laboratory (LANL) panel face size of 4, a common size. Although insufficient for the workplace, this mask offered substantial protection from the challenge aerosol and showed good fit with minimal leakage. While testers wore the mask for an hour without difficulty, we cannot comment on its utility during strenuous work or adverse environmental conditions.

2) Eye protection : Goggles, or face shields (Figure 2), should be used to prevent virus exposure of the eye mucosa. Important: goggles need to fit the user's facial features and have to be compatible with the respirator. Face shields are personal protective equipment devices that are used by many workers (e.g., medical, dental, veterinary) for protection of the facial area and associated mucous membranes (eyes, nose, mouth) from splashes, sprays, and spatter of body fluids. Face shields are generally not used alone, but in conjunction with other protective equipment and are therefore classified as adjunctive personal protective equipment.

We, at our centre, Calcutta National Medical College designed a simple, easy to make, yet effective disposable head gear, 10 of which can be made under 0.5\$. Fig (2a,b,c) shows the model "A" face shield head gear prototype used.

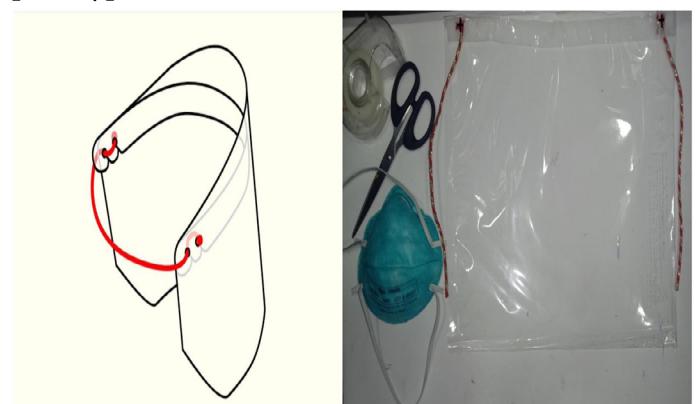


Figure 2: a) Prototype Design b) Material used in making face shield.



Figure 2c: Showing face shield application over N95 mask .

Advantages and disadvantages of face shields compared with other forms of face/eye protection that were noted by us are enumerated below (i.e., protective facemasks [filtering facepiece respirators, medical/surgical masks], goggles, safety glasses)in Table 2^{7,8,9}.

TABLE 2:

Advantages and disadvantages of face shields compared with other forms of face/eye protection

S.no.	Advantages	Disadvantages
1	● more comfortable	● fogging
2	● protect a larger portion of the face	● bulkier than goggles and safety glasses
3	● easy to don and doff	● optically imperfect
4	● less fogging than goggles	● some models may not fit properly over some respirators (e.g., duckbill filtering facepiece respirators)
5	● can be worn concurrent to other face/eye PPE	● peripheral fit poorer than protective facemasks
6	● no impact on breathing resistance	● glare
7	● no fit testing required	

There is evolving evidence that otolaryngologists are among the highest risk groups when performing upper airway surgeries and examinations. Emergency procedures like tracheostomy, cut throat injuries etc. are some unavoidable short duration exposures, but carry a high risk of transmission of COVID-19 for on duty resident surgeons. We devised a better face shield which will provide a 360 degree protection from any aerosol exposure during the operative procedure . This can be worn over N95 respirators after sterilization with 1% hypochlorite or surgical spirit & discarded after single use. As these procedures last for a shorter duration , the little discomfort caused by wearing is tolerable in view of the protection it provides.

Its use can be widened to other high risk procedures like oropharyngeal (throat) swab sample collection, emergency intubations & many more.

The fig3(a,b) shows the model “B” head gear prototype worn by resident surgeon.



Figure 3: a) Showing 360 degree protective head gear , b) Head gear used by resident surgeon (to be worn over N95 mask & PPE gown)

A 12 cm x 8 cm rectangular opening is created at the bottom of the face shield, near the supraclavicular region. Surgical mask is taped over the opening from inside. It will serve as a route for air entry, and will also prevent entry of any aerosol generated during the procedure. This model provides a better protection in procedures associated with high amounts of aerosol generation & thus belonging to the high risk category. The gear is for one time used & are disposed of according to the biomedical waste management plan in yellow bags & containers.

Upon researching how the problem of scarcity in PPE is being tackled in different parts of the world, we found many similar projects being practiced. In American Architects and Universities Leverage Open-Source Design¹⁰ , using a file created by Erik Cederberg (of Swedish manufacturer 3DVerkstan), a host of American architecture studios (BIG, Brooks + Scarpa, Höweler + Yoon, Jenny Sabin, Edg, Weiss/Manfredi, Kohn Pedersen Fox and Handel Architects,) alongside the faculty in the Architecture, Art and Planning Department of Cornell University have begun leveraging their facilities, mass producing personal protective equipment to help combat the nation-wide shortage. A majority of their output has already been donated to Weill Cornell Medical Centre.

Kitchener-based Inksmith is helping in reducing the scarcity. Though focusing primarily on STEAM (science, technology, engineering, arts and math) learning through such methods as 3D printing¹⁰.

Inksmith has pivoted to mass producing Health Canada –certified face shields

Originally able to manufacture only a handful at a time, the Kitchener-based company now estimates their capacity at 8,000 per day.

In the U.K, Batch.works, a design company that 3D prints fetching housewares¹⁰, is now dedicating its factory to producing face shields. Batch.works hopes to soon send out 1,000 units a day through a partnership with local delivery company Pedal Me. Scuba diving masks being used as head gear with special filter attachment at the air inlet at top. The scuba diving masks and snorkeling goggles are available on Amazon and produced by companies such as sports giant Decathlon, while Acosta's team has made their design freely available, and is encouraging those with 3D printers to create the attachments and give them to hospitals which are lacking protective equipment.

The masks, however, have yet to be quality tested in a lab.

The project has gained traction with volunteers based in countries such as Colombia, Chile, the USA, UK, Australia, Germany, Canada, according to Acosta, printing the devices or solving logistical issues.

There is currently no universal standard for face/eye protection from biological hazards¹¹. Therefore, the recommendations for the proper use of face shields vary widely, indicating the need for a consensus on the use of certain face/eye protection for specific medical procedures¹².

OSHA's Bloodborne Pathogens standard (1910:1030 subpart (d)(3)(i)) states: "Masks in combination with eye protection devices, such as goggles or glasses with solid side shields, or chin-length face shields, shall be worn whenever splashes, spray, spatter, or droplets of blood or other potentially infectious materials may be generated and eye, nose, or mouth contamination can be reasonably anticipated."¹³.

Conclusion:

With over 100 countries worldwide now dealing with the impacts of COVID-19, significant pressure has been placed on critical medical supplies (masks, gear, ventilators etc), healthcare facilities and hospital staff. In response, a host of international designers, manufacturers, makers and producers have leveraged their collective resources to offer innovative solutions that alleviate the strains on medical equipment as well as to ensure front line workers stay safe. Our attempt was to put forward the world a way to fight the scarcity & discuss some of the most striking initiatives being taken in this direction. Popular news outlets report unconventional solutions for PPE at local hospitals, such as plastic garbage bags for gowns and plastic water bottle cutouts for eye protection¹⁴.

Plans for resupply through the repurposing of industrial capacity and other means are welcome but seem unlikely to solve the shortage quickly enough as supply chains become more dysfunctional in the pandemic. The increase in people with respiratory symptoms visiting clinics and needing care is straining our healthcare system's supply of PPE in many areas. At this hour, these innovations provide much needed respite to healthcare workers and are in fact the need of the hour, until adequate PPE is available to the hospitals.

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PMMC FLAP FOR HEAD & NECK RECONSTRUCTION –A TERTIARY CARE CENTRE EXPERIENCE

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Abstract:

Objectives: The Pectoralis Major Myocutaneous Flap is a reliable regional flap for reconstruction in head and neck malignancies. The study is about our experience with PMMC flap reconstruction at our institution.

Materials: We studied 9 cases of Head and neck malignancies where resection was possible and needed reconstruction. The study was conducted in the department of ENT, Head & Neck surgery at Calcutta National Medical College & Hospital, Kolkata, over a period of 12 months, February 2019 to January 2020.

Result: Majority of the patients presented at locally advanced stage of the disease (67%) with male: female ratio of 2:1, mainly in their 5th -6th decades of life. All our patients had SCC of the oral cavity, under the subsets of buccal mucosa, lower alveolus, tongue and FOM in the decreasing order. Main complication met was orocutaneous fistula (22%) followed by skin/partial flap necrosis and wound infection (11% each). One patient had recurrence.

Conclusion: PMMC flap is still a first choice for reconstruction of head and neck malignancies, in places with limited resource and expertise.

Keywords: Head and neck reconstruction, head and neck malignancies, Pectoralis major myocutaneous flap, PMMC flap.

Introduction:

More than 50% of head and neck malignancies in the world occurs in Asia, especially India. Head and neck cancers accounts for 30% of cancers in India, where 60 – 80% patients present at advanced stages requiring a composite resection¹. The cancers ranging from carcinoma lower and upper alveolus, buccal mucosa, tongue, floor of mouth, gingivobuccal sulcus and mandible etc. The most common histological variant is squamous cell carcinoma². According to the statistics, globally the number of new cases of lip and oral cavity cancer is 3,54,864 and the number of deaths in the same year is 1,77,384³. The gold standard for reconstruction in head and neck cancers is microvascular free flap, which requires resources and expertise. In advanced cases where composite resection is required the principal mode of reconstruction is the PMMC flap. We present a prospective analysis of 9 cases of PMMC flap reconstruction done at our institution.

Materials & Methods:

A total of 9 cases of PMMC flap reconstructions were done over a period of 12 months, February 2019 to January 2020, at the Department of ENT, Head & Neck Surgery, CNMCH, Kolkata. After a detailed clinical history, examination and relevant investigations, the cancers were staged and operated respectively and reconstruction was made with ipsilateral PMMC flap. Procedures were performed as per standard protocol. Informed consents were obtained from all the patients. Patients who underwent primary closure and reconstructed with other flaps were excluded from the study.

Technique of harvesting PMMC flap:

- The surface marking of the vascular pedicle was made by drawing a line from the ipsilateral acromion to the xiphisternum and another line vertically from the mid-point of clavicle intersecting the first line (Ariyan Line).

A skin island was positioned over the pectoralis muscle along the course of this vascular pedicle.

- The skin island of appropriate size was designed as an ellipse with a transverse axis over the 5th rib medial to the nipple or in the inframammary fold.
- The skin and breast tissue above the skin island was widely elevated from the pectoralis major muscle upto the clavicle (Fig.1)



Figure 1

- Then skin island was incised in a beveling fashion to incise as many myocutaneous perforators as possible and elevated cephalad in the retropectoral area between the pectoralis major and minor muscle plane, along the lateral border of the pectoralis major muscle. Pectoral branch identified on the undersurface and keeping it in view further division of the flap was done (Fig. 2)



Figure 2

In case of osteomyocutaneous flap, usually 5th rib is chosen and dissected out by dividing the intercostal muscles and gently dissecting away the pleura from the undersurface of the rib. Rib was then divided using rib cutters as per the defect.

- The humeral and sternal attachments were divided for transposition into the neck and clavicular attachments and medial and lateral pectoral nerve were divided for adequate arc of rotation and to avoid supra-clavicular hump.

- The flap was then mobilized into the neck, forming a tunnel at the subplatysmal plane above the clavicle without any compression (Fig.3)

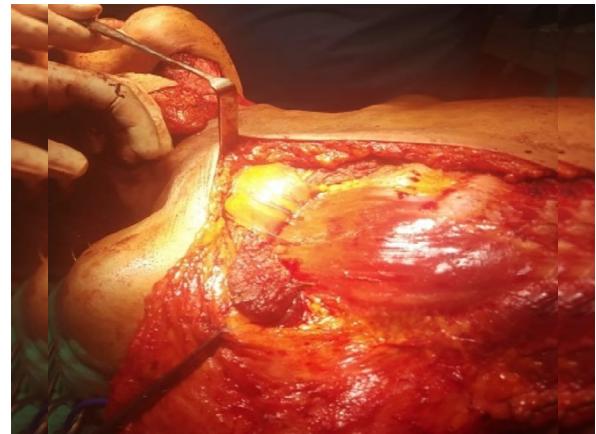


Figure 3

- Then with 3.0 vicryl the flap was sutured with the defect (Fig.4).



- Suction drains placed and wound closed in layers. Donor site closed primarily (Fig.5)



Figure 5

Observation & Results:

A total of 9 case was analyzed, of which 6 were male and 3 were female with a male: female ratio of 2:1. The age ranged from 38 to 70 years with the majority falling in the 5th & 6th decades. All the patients in our series had squamous cell carcinoma of the oral cavity, with majority under the subset buccal mucosa making up to 44%, followed by lower alveolus and tongue making up to 22% each.

Table 1: Patient's Demographic details (n=9).

Mean Age (years)	54 (Range: 38-70)	
Sex	No. of patients	Percentage
Male	6	67%
Female	3	33%
Primary Site	Percentage	Percentage
Buccal Mucosa	4	44%
Lower Alveolus	2	22%
Tongue	2	22%
Floor of Mouth	1	11%

One patient had carcinoma of floor of mouth (Table 1).

Out of 9, 6 patients had moderately advanced disease (Fig.6) with varying involvement of the gingivobuccal sulcus, retromolar trigone, mandible and skin.

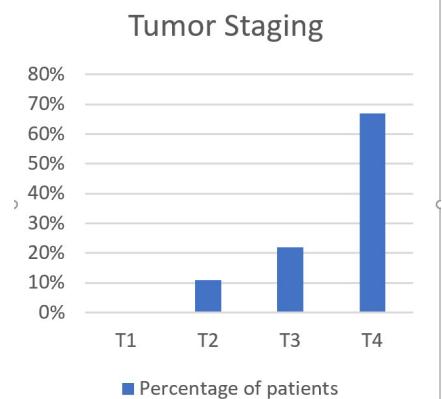


Figure 6: showing distribution of patients of different stages.

All patient underwent wide local excision of the primary tumor with modified radical neck dissection. 5 patients underwent segmental/hemi mandibulectomy along with the primary resection.

All the patients underwent ipsilateral PMMC flap reconstruction including one osteomyocutaneous flap.

The average duration of the hospital stay was 12 days (7-40 days).

During the perioperative period the most common complaints were pain followed by trismus and dysphagia. Of the 9 flaps, 3 had complete uptake of flap and the rest had complications but without total flap failure.

Among the complications met which were all flap related, orocutaneous fistula was the commonest followed by skin/partial flap necrosis and wound infection (Table 2)

One patient had recurrence of the primary after a period of 6 months. All the complications were managed conservatively.

Table 2: Post-Operative Complications:

Complication	No. of Patients
Skin Flap necrosis	1
Partial Flap Necrosis	1
Wound Infection	1
Orocutaneous Fistula	2
Total Flap Failure	0
Recurrence	1

Discussion:

India has a high mortality rate in Head and Neck cancers due to its late presentation⁴. Majority of the patients in our series (67%) was in locally advanced stage. This leads to a composite resection and therefore requirement of reconstruction. In reconstruction the selection of flaps is according to the reliability in providing coverage, restoring contour and function, without compromising the primary aim of adequate tumor resection.

The use of muscle as a flap requires movement of a part or all of the muscle, without interrupting its circulation. A precise knowledge of vascular anatomy of the muscles to be utilized is essential. The well-established Mathes and Nahai classification of vascular anatomy of muscles is used for this purpose. Accordingly, the five patterns are: Type 1-One vascular pedicle; Type 2-dominant pedicle(s) and minor pedicle(s); Type 3- two dominant pedicles; Type 4- Segmental vascular pedicles; Type 5-One dominant pedicle and secondary segmental pedicles. PMMC flap was first described by Stephen Aryan in 1979⁵.

Surgical Anatomy:

Pectoralis major muscle originates from the medial half of clavicle, medial part of sternum and first 6 ribs and inserts on the lateral lip of the bicipital groove of the humerus. It falls under type 5 pattern, the dominant vascular pedicle being pectoral branch of thoracoacromial artery, branch of subclavian artery and the segmental secondary pedicles being perforators of internal mammary artery from 1st to 6th intercostal spaces. So, the entire muscle would survive elevation on either vascular system. The arc of rotation is wide which is oblique from the lateral end of clavicle to the xiphoid process and so it is a flap of choice for composite defects of lower face and oral cavity. The skin territory is located between the nipple and the sternum over the 5th- 6th ribs. A variation of the PMMC flap by placing the skin unit more distally in the inframammary crease increases the arc of rotation. The vascular communication between the muscle and the periosteum of the fifth and sixth ribs allows the elevation of vascularized rib for mandibular reconstruction.

Causes of failure:

The main morbidity of any flap is failure. As with any other procedure, the error in judgement, technique and concurrent illness plays a vital role. The causes of flap failure are due to planning errors, technical errors and post-operative errors. Planning errors include, inadequacy in design, not considering vasculature in previously operated or irradiated patient and not considering arc of rotation in obese patient. The flap of choice and alternatives should be determined pre-operatively. Technical errors include, inappropriate harvesting of flap and aligning the flap to the donor site, incorrect pedicle division, inadequate tunneling, increased tension, bleeding and inadequate debridement leaving nonviable tissue leading to flap failure. Post-operative care errors include, positioning with pressure, tight dressing compromising circulation, improper drain, perioperative antibiotics and immobilization.

Complications:

The drawback of PMMC flap is the higher complication rate as seen in the study done by El-Marakby⁶. The study done by S.Sen et al shows that the success rate of PMMC flap in terms of achieving treatment goals is higher in spite of the increased complication rates⁷. Wei Wei et al in his study says that the complication rates of PMMC flap in primary reconstruction is lesser than that of secondary reconstruction but has a similar survival prognosis⁸. Our study had orocutaneous fistula (22%) as the most common complication, which was managed conservatively.

Skin/partial flap necrosis and wound infection was seen in around 11% of patients respectively, which coincides with the studies done by Atanu Bhanja et al and Anehosur et al^{9,10}. The loss of adduction and medial rotation was well tolerated by patients in our study as no patient had major complaint of dysfunction. The chest wall cosmesis was also acceptable (Fig.7).



Figure 7: Acceptable chest wall cosmesis, 2 months post operatively.

Although majority of the patients presented at the late stage and had consecutive post-op complications, the functional and cosmetic outcome was satisfactory. Despite the drawbacks, due to the ease of technique; the bulkiness leading to the usage in advanced cases; as a salvage flap in free flap failures, recurrences and laryngectomy post irradiation, PMMC is considered a versatile flap¹¹⁻¹⁶.

Conclusion:

In the age of microvascular free flap transfer, PMMC flap is still the first choice for head and neck reconstruction, at places with less expertise and facilities. In head and neck cancers constituting complex functional and aesthetic reconstruction, PMMC flap gives a favorable outcome. Due to its ease, reliability and less complications it is the workhorse flap in places with increased patient load. According to our experience which has shown a low rate of complication, PMMC flap is an excellent choice in head & neck reconstruction.

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Unresectable Tumors Of The Head And Neck

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Introduction:

Head & Neck cancers form 30 % of the total cancer burden worldwide. In India about 75% of cancer patients, like in most of the developing world , present in advanced stages. Thus proper decision making in the management of head and neck cancer cannot be over emphasized. The treatment offered is either surgery or Radiotherapy (often with chemotherapy) or more often a combination of both.

The decision for surgery is by and large dependent on the resectability of the tumor. If the tumor is unresectable, then radiotherapy or palliative care is obvious alternative. That leads us to an important question: What is an unresectable tumor? There is general consensus that tumors with AJCC T4b staging are surgically unresectable. The T4b stage emphasizes the poor prognosis unresectability. Tumors staged T4a and lower are generally considered to be resectable.

The main criteria for staging a primary head and neck tumor as T4b are as follows:

1. Vascular (carotid) encasement and invasion.
2. Prevertebral space invasion.
3. Mediastinal invasion.

However, it needs to be stressed that 'resectable' does not necessarily mean 'operable'. The 'operability' is dependent upon various patient, clinical and external factors. A decision has to be made regarding the complete resection of the tumor and what impact it will have on the patients quality of life, profession and self image thereafter. The successful resection of the whole tumor may be good for the self esteem of the surgical team but the morbidity associated with the primary surgery may give the patient a quality of life with questionable additional value.

The patients choice will be important where the loss of an organ (tongue or larynx) or a function (swallowing or speech) alongwith a significant cosmetic morbidity will give him, say a 10-15% increase in 5 year survival compared to RT or sometimes even palliative care.

Keeping this in mind it will be helpful to group patients in the following categories ⁽¹⁾:

1. Unresectable
2. Resectable with limited "collateral damage"
3. Resectable – Extensive surgery with major QOL issues.

Factors determining Operability:

By an large the factors that determine morbidity also determine the 'operability'. These include:

1. Tracheal invasion: Intraluminal invasion of the trachea and its extension into the mediastinum indicates advanced primary tumor and is associated with low survival.
2. Oesophageal invasion: Invasion of the oesophagus is associated with poor resection margins and thus poor survival because of its proximity to the prevertebral soft tissues.
3. Laryngeal cartilage penetration: The traditional belief is that involvement of the thyroid cartilage is a contraindication for RT and an indication for surgery. However, there are studies that indicate that T4 tumor patients with minimal laryngeal cartilage involvement do well with RT/CRT (2)
4. Pre-epiglottic fat involvement: May require resection of a part of the base of tongue with its associated problems regarding swallowing.
5. Dural spread.

6. Bone infiltration (mandible, maxilla and skull base): Involvement of the skull base by a malignant primary tumor or metastatic nodes (IIb) compromise oncological margins and thus adversely affect prognosis.
7. Orbital involvement: The decision to exenterate even a partially functioning eye in the name of oncological clearance has significant implications for the patient.
8. Brachial plexus involvement: Either by the primary tumor or the metastatic nodes. Will be associated with involvement of the scalene muscles and thus poor operability.
9. Co-morbid conditions: Like chronic debilitating diseases or overall poor general health of the patient which increases surgical morbidity.
10. Condition of the neck nodes: Especially extranodal extension.
11. Preference for organ preservation: The patients choice is very important.
12. Expected functional outcomes following surgery.
13. Chances of getting pathologically negative surgical margins.
14. Histological grading of the tumor.
15. Patient compliance.
16. Survival statistics: The patient needs to be informed in detail about the survival chances with each treatment modality so that he/she can make a choice balancing the QOL with survival.

Unresectability does not necessarily mean incurability:

An unresectable tumor is not necessarily incurable and definitely not untreatable. Head and neck lymphomas, nasopharyngeal and oropharyngeal SCC are good examples of such tumors. So all is not lost if the tumor is unresectable or deemed inoperable. There are other good treatment options available the decision about which should be made after detailed discussions with the patient. After all it is his/her life and his/her body. If they decide to chose QOL over a marginal increase in survival or otherwise, we should respect their judgement and go along with it.

Management Options for unresectable/ inoperable tumors:

1. Radiotherapy
2. CT + RT
3. Palliative care: In the form of 'Best supportive care'(3) which is defined as, treatment administered with the intent to maximise the QOL without a specific antineoplastic regimen. This obviously

includes all kinds of symptomatic and supportive treatment. BSC alone is associated with a median survival of 3-6 months.

Palliative care can also include palliative radiotherapy which though part of an antineoplastic regimen is not administered with curative intent.

Conclusion:

Resectable tumors are not necessarily operable. The resectability is dependent primarily on tumor-factors while the operability is dependent primarily on patient factors. However, it needs to be emphasized that unresectable doesn't necessarily mean incurable. So, the treatment of head and neck cancers needs to be individualised depending on the patients preferences after detailed discussions with them.

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ESTHESIONEUROBLASTOMA: A RARE PRESENTATION

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Abstract:

Introduction- Esthesioneuroblastoma, also known as Olfactory Neuroblastoma, arise from the basal cells within the olfactory neuroepithelium. It accounts for <5% of the sinonasal malignancies. The slow growing nature of the tumor, often delays its diagnosis. The nature of invasion into surrounding structures including Dura and middle cranial fossa, increases its morbidity.

Clinical Presentation- A 26 years lady presented with a gradually progressive swelling in the left cheek, which was initially painless but later it became painful and was associated with loosening of teeth of upper jaw. On examination, there was a hard, non tender and non pulsatile swelling, extending from left nasolabial fold to the zygomatic bone and from the orbit till the angle of mouth. It was fixed to the overlying skin, and the margins were not palpable on any side. Radiological studies (CT and MRI) showed a heterogenous SOL, involving the left side of the cheek, with extensive spread to the surrounding structures including the adjacent maxillary antrum and left side of the nasal canal, with erosion of the adjacent bony alveolus and floor of left maxillary antrum. FNAC from the swelling was suggestive of poorly differentiated malignant neoplasm. Biopsy taken from the nasal mass was diagnostic of neuroblastoma.

Surgical Management- The patient underwent left sided extended maxillectomy by Weber Ferguson's approach with a Dieffenbach's extension under general anaesthesia. Later the patient was sent for chemoradiation. The patient was followed up and after 6 months, an artificial denture was given to the patient after completion of her radiotherapy.

Discussion- Esthesioneuroblastoma is a very rare malignancy of the olfactory neuroepithelium. Certain histological features helps diagnosing this tumor. Confirmatory diagnosis requires examination under electron microscope. The staging system proposed by Kadish et al. modified by Morita is now widely accepted. This staging system is predictive of disease-related mortality.

Key words- Esthesioneuroblastoma, Olfactory, Paranasal Sinus-Nasal Cavity, Neoplasms, Head and Neck

Introduction-

Esthesioneuroblastoma, known as Olfactory Neuroblastoma, arise from the basal cells within the olfactory neuroepithelium. It consists of <5% of the sinonasal malignancies, extremely rare. It was originally described in 1924 by Berger et al¹ in the french literature 'esthesio-neuroepitheliome olfactif' with the highest series of cases being reported by Jackson et al in 1984 while highest number of cases was reviewed by Skolnick et al³ in 1966. Olfactory neuroblastoma exhibits a wide range of morphological diversity ranging an indolent growth to highly aggressive neoplasm with propensity of rapid metastasis⁴.

It has got a bimodal age distribution of 20 and 50 years and more common in females than males. Though it presents with nasal symptoms like obstruction and intermittent epistaxis, the diagnosis is often delayed due to its slow growing nature. The disease extends its morbidity via direct extension and invasion into the surrounding structure (25% of the cases invades dura and anterior cranial fossa). Radiographic features show a contrast enhancing lesion with expansile and destructive growth with expansile and destructive growth patterns. Genetic studies have shown involvement Hedgehog pathway, MYC and KDR genes implicated for it.

CLINICAL PRESENTATION:

A 26 years female presented with a swelling in the left cheek , which was progressively increasing in size. After a month, she started developing pain over the region and loosening of tooth in the left upper jaw.

It was also associated with an nasal blockage. Her symptoms have been increasing ever since. She also subsequently developed difficulty in chewing and a loss of appetite.

On examination, she had developed trismus. There was a hard ,non tender and non pulsatile swelling measuring about 8 x 9 cm in antero posterior and and craniocaudal directions. It extended from left nasolabial fold to the zygomatic bone and from the orbit till the angle of mouth. It was fixed to the overlying skin, and the margins were not palpable on any side. There were no palpable lymph nodes.



Figure 1



Figure 2

Figure 1 and Figure 2 showing clinical presentation of the patient.

Radiology: CT scan reveals opacity in the sinuses, more marked on the left side and a left upper alveolobuccal mass with erosion of the adjacent bony alveolus and floor of left maxillary antrum.

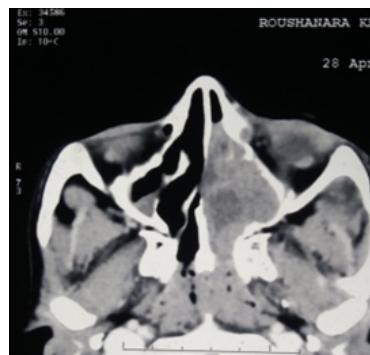


Figure 3



Figure 4

Figure 3 and 4 showing CT scan findings.

MRI (Plain and contrast) are suggestive of large, heterogenous, contrast enhancing soft tissue mass seen involving the left side of the cheek, with extensive spread to the surrounding structures including the adjacent maxillary antrum and left side of the nasal canal.



Figure 5

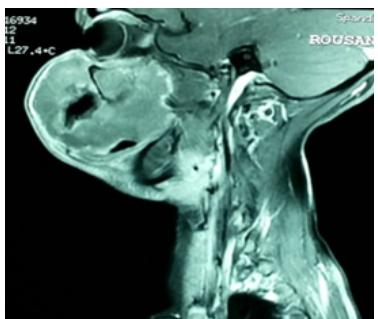


Figure 6

Figure 5 and 6 showing MRI Findings.

Histopathology- FNAC from the swelling was suggestive of poorly differentiated malignant neoplasm. Biopsy taken from the nasal mass was diagnostic of neuroblastoma. The histopathology was confirmed from two different centres and the report was similar from both

SURGICAL PLAN:

The patient was planned for a total left sided extended maxillectomy under general anaesthesia. Weber Ferguson's incision with a Dieffenbach's extension was given and cheek flap elevated. Inferior orbital nerve divided. Frontal process of maxilla and the lacrimal bone,floor of the orbit, zygomatic process and the palatae were divided and osteotomy to separate maxillary tuberosity from the pterygoid plates

Involved parts of pterygoid and masseter were excised. A part of the skin with subcutaneous involvement was also excised.

Bleeding from the internal maxillary artery was controlled by ligation and oozing from the pterygoid plexus by packing.

Dacrocystorhinostomy was performed, where the sac was identified and marsupialised.

The skin flap was closed in layers after putting an occluder in the guttaparcha.

Following extended maxillectomy, the patient was referred to the department of Radiotherapy for further treatment.

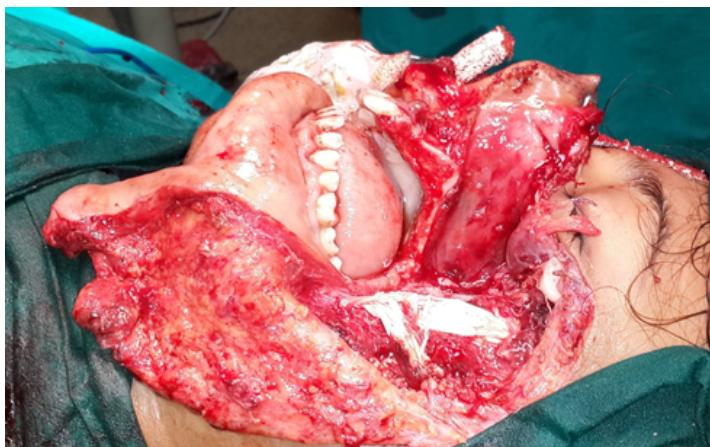


Figure 7 :Intraoperative picture of the patient after removal of the mass.



Figure 8



Figure 9

Figure 8 and Figure 9 showing Post operative outcome.

DISCUSSION:

Esthesioneuroblastoma is a very rare malignancy of the neuroepithelium. Embryologically, the olfactory nerves develop from the olfactory placode present in the fetal olfactory mucosa^{5,6}. Histologically, there are a number of criterias that help in its diagnosis: neuroepithelial cells arranged in the classic pseudorosette pattern; fibrillar intracellular background; marked microvascularity; and round or fusiform cells approaching the size of lymphocytes with poorly defined,

almost nonexistent cytoplasm^{7,8,9}.

Correct diagnosis often requires confirmatory examination with electron microscopy for the detection of neurosecretory granules. More recently immunohistochemical methods for detection of neuronspecific enolase (NSE) and S-100 protein with negative epithelial, muscle, and lymphoid antigens allowed further confirmation of ENB^{7,10}.

The staging system presented by Kadish et al.¹¹ in 1976 based on tumor extension, and modified by Morita¹² has been widely accepted. This staging system is predictive of disease-related mortality. , as our own patient.

It is a slow growing tumor, and patient generally presents late due to non specific symptoms and very rarely presents with a facial swelling.

Our present presented with the facial swelling as her chief complain with other non specific complains of frequent epistaxis and nasal obstruction.

Involvement of the maxilla before involvement of the orbits or the anterior cranial fossa is rare.

An extended maxillectomy was done for the patient. A skin grafting was avoided due to uncertainty of spread. The patient was followed up and after 6 months, an artificial denture was given to the patient after completion of her radiotherapy.

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UNSAFE CSOM WITH POST-AURICULAR ABSCESS IN PATIENTS WITH MICROTIA: OUR EXPERIENCE

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Abstract:

Microtia is a developmental anomaly of ear due to failure of fusion of hillocks developing from 1st & 2nd pharyngeal arches. Microtia occurs once in every 6000 births. In 10% of cases, the problem is bilateral. The right side is more commonly affected than the left and males are more often affected than females. Incidence of atresia of external auditory canal to be 0.5 per 1,00,000 population, incidence of membranous atresia being 20 times less than the solid form. Congenital aural stenosis as compared to aural atresia, carries a much greater risk of cholesteatoma. Jahrsdoerfer & cole reviewed 600 cases of major congenital ear malformation. Fifty patients (54 ears) were found to have aural stenosis. The most significant finding was that in children of 12 years or older with a meatus narrower than 2mm, 91% develop cholesteatoma. Here, we will describe 2 cases with congenital anomaly of external ear, presenting with recurrent abscess in post aural region. Case one was having unilateral microtia with atresia of external auditory canal whereas case two was having bilateral microtia with stenosis of bilateral external auditory canal. Both of cases were operated and reconstructive procedures were done for better audiological as well as better cosmetic outcome and disease free ear.

Key words- Congenital Microtia, Ear Canal, Constriction, Pathologic, Cholesteatoma, Abscess

Introduction:

A study by becker & tos¹ showed the incidence of atresia of external auditory canal to be 0.5 per 1,00,000 population, incidence of membranous atresia being 20 times less than the solid form.

Congenital aural stenosis as compared to aural atresia, carries a much greater risk of cholesteatoma.

Jahrsdoerfer & cole² reviewed 600 cases of major congenital ear malformation. Fifty patients (54 ears) were found to have aural stenosis.

The most significant finding was that in children of 12 years or older with a meatus narrower than 2mm, 91% develop cholesteatoma.

Microtia is the developmental anomaly of ear due to failure of fusion of hillocks developing from 1st & 2nd pharyngeal arches. Microtia occurs once in every 6000 births. In 10% of cases, the problem is bilateral. The right side is more commonly affected than the left and males are more often affected than females.

The normal size of the auricle at birth is 66% of the length & 76% of the width of an adult ear. By the age of six, the auricle has attained 90% of adult proportion³.

Case 1:

A 7 year old male child was brought to the out patient department with complain of right sided post auricular swelling and pain since last 2 days.

On Examination - There was a globular post-auricular swelling on the right side of size 2x2 cm², fluctuant in nature, tender to touch with tense, inflamed and excoriated overlying skin having pus points.

Local examination further revealed : the child had right sided microtia with atresia of external auditory canal. External ear consisted of lobule, tragus and a small bulge of cartilagenous tissue behind an atretic canal.

However, lobule and tragus of the diseased ear were present almost at the same level compared to the other side. His left ear was completely normal.



Figure 1 : Photograph at initial presentation

On further enquiry, the parents gave a history of similar post-auricular abscess of same ear , which ruptured and healed spontaneously without any treatment.

Blood investigations revealed raised wbc count ($18.2 \times 10^3/\text{ml}$), predominantly polymorphs. Other parameters were within normal limit

Microscopic examination of the pus from post-auricular abscess showed gram positive cocci in cluster with no growth of pathogenic organism on culture. BERA : bc-abr suggestive of cochlear pathology in right ear. Ac-abr could not be done due to external ear pathology. Ac-abr of left ear suggestive of minimal hearing loss.

Pure tone audiometry (PTA) & tuning fork tests gave inconsistent results.

High resolution CT (HRCT) temporal bone showed right sided post-auricular thickening with soft tissue & fluid, along with thickening of pinna. Eac showed fluid & granulation with circumscribed cystic lesion in middle ear. Ossicles were clear, Internal acoustic canal (IAC) was normal.

Operative findings: in order to locate the antrum of the diseased ear the mastoid tip of that side is palpated and its position is compared with that of the opposite side. Bilaterally the mastoid tip & the tragus were lying at the same level. Lobule was almost 2cm above & anterior to the mastoid tip. Eac was probed and found to be blind measuring 3mm

A curvilinear incision was given passing over the post-auricular abscess upto 1cm above the mastoid tip running behind the lobule. After initial dissection spine of henle was identified which was rudimentary. Mac ewan's triangle was not well defined. Mastoid antrum & attic were hypoplastic. Cholesteatoma was seen in antrum, sinodural angle, attic and going further anteriorly into the eustachian tube opening. Eustachian tube orifice provided an important landmark to identify the otherwise malformed middle ear. Cholesteatoma sac was found lying on the vertical segment of the facial nerve. Dura was low lying. Incus and malleus were malformed & fused together. Canaloplasty done. Meatoaplasty was done by pulling and suturing the tragus more anteriorly & lobule further inferiorly. A stent was kept to maintain the patency of the eac.



Figure 2 : Post operative photograph

Case 2:

A 17 year old female was brought to ent opd with complain of left post auricular swelling with pain and left ear discharge since last 5 days. She also complains of left sided facial asymmetry and decreased hearing from the same side since childhood.

On physical examination, the patient had bilateral microtia with a diffuse swelling over left post-auricular area extending upto the angle of mandible and lateral aspect of upper third of neck. The swelling was fluctuant and tender to touch.

On left side, there were two cartilagenous spicules in place of the lobule lying inferiorly to the meatus and an isolated tag of soft tissue almost 2 cm posterior to the eac. Tragus was rudimentary. Left external auditory canal was stenosed & filled with foul smelling whitish discharge. Bilateral eac were placed more posteriorly & at a lower level, almost 3 cm below the level of the horizontal line drawn from the lateral canthus of eye on the temporal bone.



Figure 3 : Pre operative photo of case 2

Past history of 2 similar episodes were present 4 yrs & 8 yrs ago for which the patient underwent incision and drainage.

Blood investigations revealed raised wbc count ($14.1 \times 10^3/\text{ml}$), with other parameters being within normal physiological range.

Microscopic examination of pus from the abscess showed gram positive cocci but without growth of any organism.

Audiological evaluation: Pure tone audiometry showed 55db conductive hearing loss in left ear. Bone conduction masking could not be done as the patient is having postauricular pain.

Right ear- Normal hearing, bone conduction threshold within normal limit.

Tympanometry & Otoacoustic Emission (OAE) could not be done due to active discharge.

Tuning fork test :

	Pre op		
	256hz	512hz	1024hz
Rinne:	+/NP	+/NP	+/NP
Weber :	----->		
ABC :	-----Eq/NP-----		

[NP: Not perceived, Eq: Equivocal, + : Positive]

HRCT temporal bone: homogenously mildly enhancing soft tissue density seen infiltrating the left external and middle ear cavities with erosion of posterior & inferior bony wall of eac, scutum & long process of incus. It is seen completely filling epi, meso & hypo tympanum. It is also extending in prussack's space, aditus & mastoid air cells with their erosion & opacification. It is seen eroding the lateral semicircular canal with patchy dehiscence of facial canal. Evidence of heterogeneously enhancing thick walled collection is noted in left posteroinferior auricular region.

Operative findings: probing revealed a stenosed eac running downward & posteriorly. An initial small incision of 2cm was given just below and almost 1.5cm posterior to the cartilagenous remnant away from the mastoid tip to drain the abscess. The incision was extended upward and anteriorly running between the soft tissue tag and the eac, upto 1.5cm above the meatus. Spine of henle was rudimentary. Drilling revealed sclerosed mastoid antrum. Granulation tissue present in aditus, attic & middle ear. Malleus & incus were malformed & fused; stapes head & stapedius tendon were visualised. Round window found to be placed anteriorly. Lateral semicircular canal was eroded. Abscess found to extend from eac inferiorly. Canaloplasty was done. The soft tissue tag was pulled & sutured superior to the meatus. Meatoplasty was maintained by keeping a stent.



Figure 4 : Post operative photo of case 2

Tuning fork test :

	Post op		
	256hz	512hz	1024hz
Rinne:	+/-	+/-	+/-
Weber :	----->		
ABC :	-----Eq/Eq-----		

[NP: Not perceived, Eq: Equivocal, + : Positive]

Discussion:

The auricle or pinna develops from a series of small cartilaginous tubercles or Hillocks. Hillock 1-3 comes from 1st or mandibular arch & 4-6 comes from 2nd or hyoid arch. According to park⁴, hillock 1 produces the anterior portion of the ear lobule, hillock 2 tragus & hillock 3 the ascending helix. Of the 2nd arch hillocks, 4 & 5 produces antihelix & helix, with 6 contributing to the posterior lobule.

By the end of fifth week, five branchial arches are discernable. In a 38 day old embryo, six hillocks have developed in the mesenchymal tissue of the first (mandibular) and second (hyoid) arch and a process of fusion produces a primitive ear in the 50 day old embryo.

Both case 1 & 2 presented with microtia, case 1 unilateral where as case 2 bilateral. [malformation, such as anotia and microtia, are likely to be caused by the disturbance of development at seven or eight weeks gestational age.] In both the cases tragus & lobule (1st arch structures) though present, 2nd arch anomaly was more pronounced.

The ear initially forms in the neck region and moves upward onto the head by week 10. In case 2 this migration was affected & bilateral ears were low set.

Case1 had atresia of right external auditory canal where as case 2 presented with stenosis of bilateral canal.

At 28 weeks, a core of ectoderm canalizes from medial to lateral and eventually breaks through to communicate with the conchal depression. Failure of canalization or more likely lack of ectodermal migration can lead to atresia of external auditory meatus and partial canalization leads to meatal stenosis (diameter of the canal less than 4mm).

In both the cases malleus & incus were malformed and fused. The head of the malleus and body and short process of the incus develops from meckel's cartilage (first arch derivative), whereas, the manubrium of the malleus, long process of the incus and stapes suprastructure arise from reichert's cartilage (second arch derivative). The process begins at 4 weeks & adult shape, size & ossification is present by 25 weeks.

The full-sized outline of membranous labyrinth is formed by 25 weeks of gestation.

The scc starts to develop at 35 days. Cochlea is also formed by 25 weeks.

The organ of corti starts developing as a single block of heaped up ectodermal cells at about 11 weeks. Within this mass develop inner & outer hair cells & then specialized supporting cells.

Recent studies using high resolution computed tomography suggest a higher rate of inner ear congenital anomalies affecting between 10 and 47 percent of patient with atresia.

BERA of case 1 is suggestive of cochlear pathology in right ear.

Conclusion:

Abscess are the presenting feature of congenital ear atresia. The age varies depending on deformity. So prevention is to perform reconstruction as soon as guidelines indicate in order to prevent any complication. We should be alert for bony meatal, middle and inner ear abnormality during surgery. The diseased ear are the ear with altered anatomy. Hence we will have to anticipate them. As far as hearing is concerned, restoration should be done. Staged cosmetic correction is preferred.

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ANGIOINVASIVE RHINO-CEREBRAL ASPERGILLOSION: A CASE REPORT & REVIEW OF LITERATURE

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Abstract

Background: Aspergillus spp are ubiquitous fungi. Although inhalation of its infectious conidia is common, tissue invasion is uncommon. Invasive aspergillosis is a highly lethal opportunistic infection that occurs most frequently in the setting of immunosuppression and is often rapidly progressive and fatal. Angioinvasion is a rare manifestation of disseminated aspergillosis which may result in stroke in immunocompromised individuals. Reports of such disease in patients with diabetes mellitus are rare. Rarely, aspergillosis may also occur in immunocompetent individuals.

Case Report: A 45 years old female patient, presented with proptosis, nasal obstruction and later monoparesis on right side. Nasal endoscopy showed a blackish colored mass in the right osteo-meatal complex area. Initial HPE report from that area showed granulomatous lesion suggestive of Aspergillosis. CT showed an irregular, heterogeneously enhancing mass lesion in the ethmoidal sinuses extending superiorly to the right frontal lobe destroying the floor of anterior cranial fossa. MRI showed a large area of intense, patchily enhancing, infiltrating, destructive lesion in the right basi-frontal region. Patient was started on Voriconazole. However she developed monoparesis on the right side after starting the therapy. MRI scan of brain revealed multiple, focal, altered signal intensity lesions with strong restricted diffusion in the left parafalcine region suggestive of acute cerebral infarction in the left anterior cerebral artery (ACA) territory. After initial medical management of the monoparesis, surgical debridement of the mass was done to reduce the pressure effect. Culture report was also suggestive of Aspergillosis. The patient was continued on Voriconazole therapy, but, unfortunately she succumbed to her illness despite treatment.

Conclusion: Angio-invasion in this case is most likely responsible for the monoparesis, which is uncommon in a patient with diabetes mellitus. Complete excision of the mass was not possible in this case as differentiation between brain tissue and fungal mass was not possible in this case. Voriconazole is preferred over Amphotericin-B due to its lesser toxic side-effects.

Key words: Aspergillosis, rhino-cerebral, cerebral infarctions, Aspergillus fumigatus, immunocompetent

INTRODUCTION:

Aspergillus is a ubiquitous fungus and is the most common fungal pathogen in sinus disease. Spores are also ubiquitous, usually introduced by inhalation, and are frequent inhabitants of the human upper respiratory tract¹. Aspergillus spp. Rank closely behind Candida spp. in causing invasive fungal infections in humans². Aspergillus species is pathogenic in birds and mammals. 8 out of 350 aspergillus species is associated with human disease³. Route of infection is either gastro-intestinal or respiratory tract⁴.

Invasive rhino-cerebral aspergillosis is a fatal disease, with a high mortality rate of 85%–100% despite antifungal treatment⁵. It occurs in 10%–15% of patients with disseminated aspergillosis^{6,7}. Patients at risk for angioinvasion include those with prolonged neutropenia, diabetes mellitus, recipients of haematopoietic stem cell transplants or solid-organ transplants, advanced AIDS, chronic granulomatous disease and autoimmune disorders treated with intensive immunosuppressive regimens⁸. It is, however, rarely reported in patients with diabetes mellitus.

The unique ability of the fungus to digest elastin within the vessel wall makes it highly angioinvasive, leading to a wide spectrum of neurological sequelae^{6,7}. Two factors play an important role in the pathogenesis of invasive aspergillosis:

- i) secretion of toxic substance by fungi that penetrate tissue
- ii) tissue necrosis induced by immune mechanism⁹.

Amphotericin B is the one of the commonly available anti-fungal treatments for aspergillosis and has an efficacy rate of about 40% to 60% in invasive aspergillosis^{10,11}. Amphotericin-B has considerable renal and hepatic toxicity¹⁰. Voriconazole has the advantage of better tolerance, increased efficacy, and significantly less toxicity when compared with Amphotericin B hence, is recommended as the first line treatment for invasive aspergillosis¹¹.

CASE REPORT:

A 45 years old female patient presented to the Otorhinolaryngology out-patient department with the complaints of a right sided proptosis, nasal obstruction and deterioration of vision on the same side for the last 10 months. The proptosis was insidious in onset and gradually progressive in nature. The visual deterioration gradually progressed to loss of vision. There was no associated history of epistaxis. She had diabetes mellitus but no other associated co-morbidities like cancer, AIDS, prolonged steroid intake or chronic systemic illness.

On examination there was inadequate closure of the eyelid with corneal keratinization as well as ulceration. The upper eyelid had a discharging sinus over its supero-nasal aspect. Perception of light and projection of rays was absent. Anterior rhinoscopy was suggestive of chronic rhinosinusitis. Thereafter, the patient was subjected to diagnostic nasal endoscopy which showed an irregular heterogeneous mass filling the entire ethmoidal complex area.



Fig 1: Clinical Photograph at presentation

CT scan of brain and orbit shows, irregular, heterogeneously enhancing mass lesion in ethmoidal sinuses extending superiorly to right frontal lobe destroying the floor of anterior cranial fossa with surrounding white matter edema showing mass effect. The mass extends into extraconal right orbit with the destruction of medial wall of right orbit and to the right maxillary and frontal air sinuses.

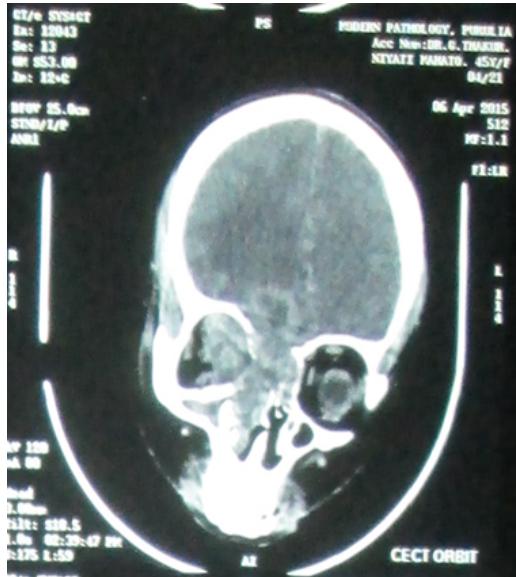


Fig 2: CT Brain and Orbit

MRI of brain in the same case showed a large area of intense, patchily enhancing, infiltrating, destructive lesion in the right basi-frontal region predominantly involving the ethmoidal sinus, nasal cavity, medial wall of the right orbit and right maxillary antrum with intraorbital and intracranial extension to the right orbito-frontal, cortical and subcortical region



Fig 3: MRI Brain and Orbit

Endoscopic biopsy provisionally diagnosed it to be a case of rhino-cerebral Aspergillosis. The hyphal morphology resembles that of *Aspergillus fumigatus*.

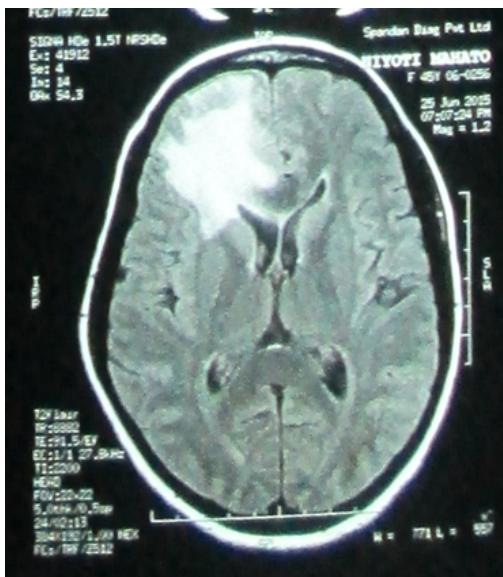


Fig 4: MRI Brain

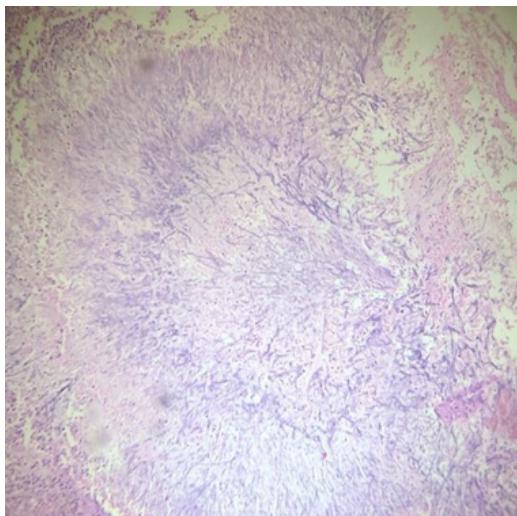


Figure 5: Slide shows hyphal elements from the material debrided from the patient (Haematoxylin-Eosin stain).

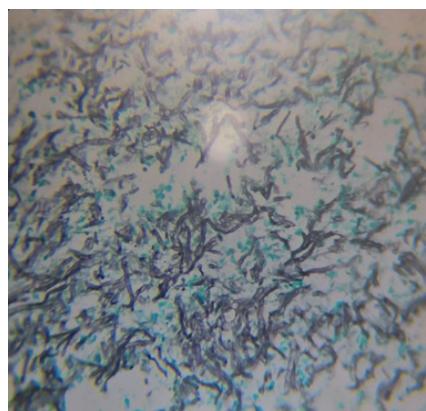


Figure 6: Slide shows hyphal elements from the material debrided from the patient (Gomori-Methanamine Silver stain).

The patient was thereafter started on Voriconazole. One week after admission and starting of medical treatment, the patient developed sudden onset right lower leg monoaresis with urinary incontinence.

There was no associated seizure, unconsciousness or vomiting.

On examination, patient was conscious, alert and co-operative. Power in the right lower leg was 1/5 while in the rest of the limbs it was 5/5. Babinski sign was positive in the right lower limb.

MRI scan of brain revealed multiple, focal, altered signal intensity lesions with strong restricted diffusion in the left parafalcine region suggestive of acute cerebral infarction in the left anterior cerebral artery territory.

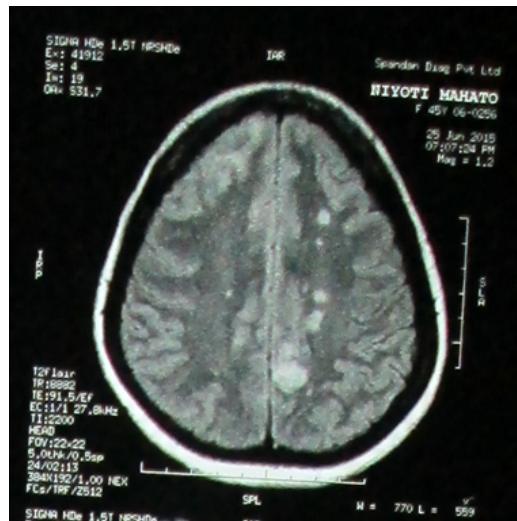


Fig 7: MRI Brain after onset of CNS Features

Neurological consultation was sought and the patient was started on intravenous mannitol and phenytoin. After stabilizing her condition, debulking of the space occupying lesion with orbital exenteration was done. The fungal mass and frontal lobe tissue was indistinctable and inseparable in places; hence complete excision was not possible.

Post-operatively specimen was sent for histopathology, KOH mount and fungal culture. HPE shows chronic granulomatous reaction. KOH mount shows hyaline, septate branching hyphae. Fungal culture morphologically resembles *Aspergillus fumigatus*.

DISCUSSION:

Since its first description as an opportunistic infection in 1953, there has been a significant increase in the number of Aspergillosis, furthermore in the number of invasive aspergillosis⁵. They are encountered more frequently in patients, whose host defense mechanisms have been compromised due to disease or due to immune-suppression and immune-modulation used for managing malignancy, autoimmune diseases and organ transplantation¹². Cases of CNS mycoses in apparently immunocompetent and healthy individuals have also been reported, mostly from India¹³. Rhino /sino-orbito-cerebral mycosis is a disease entity most often caused by the saprophytic molds *aspergillus* and *mucorales*¹⁴.

Invasive aspergillosis occurs in 10%–15% of all cases with the disseminated disease⁶.

Involvement of the central nervous system by Aspergillus spp. had been recognized as early as 1930s, and is usually a result of haematogeneous spread from another primary source such as the lungs or the gastrointestinal tract⁵⁻⁷. The unique ability of the fungus to digest elastin within the vessel walls makes it a highly virulent and angioinvasive organism^{6,7}.

There has been little data on cerebral invasive aspergillosis in patients with diabetes mellitus, thus making our patient an atypical subject. As the chest radiograph of our patient was normal, the fungal dissemination in this patient must have occurred from the paranasal sinuses. This is unusual as cerebral invasive aspergillosis is most commonly haematogeneously spread from the lungs or the gastrointestinal tract, rather than as a direct extension of sinonasal disease^{5,7}.

Aspergillosis causes an infective vasculopathy leading initially to acute infarction or hemorrhage and later into surrounding tissue as infectious cerebritis which may later evolve into an abscess⁵⁻⁷. It has an affinity for perforator arteries, and as they have a narrower lumen they tend to be affected much earlier thus affecting basal nuclei, thalamus, corpus callosum and midbrain⁵⁻⁷. Aspergillus hyphae may grow through the vessel wall leading to the formation of mycotic aneurysms of the larger arteries which may rupture causing massive hemorrhage⁵⁻⁷. Additionally vessel lumen may be completely occluded by hyphal elements leading to ischaemic stroke.

The prognosis of cerebral angioinvasive aspergillosis is poor, with a high mortality rate of 85%–100% despite aggressive therapy^{5,15}. Based on the largest randomized controlled trial, it is recommended that voriconazole be used for the primary treatment of invasive aspergillosis¹⁶, as voriconazole has the advantage of better tolerance, increased efficacy and significantly less toxicity compared to amphotericin B. It also shows less nephrotoxicity and better compliance than with amphotericin B in clinical practice¹⁶.

CONCLUSION:

Rhino-cerebral aspergillosis is a rare fungal infection. Angioinvasion occurs rarely in a case of rhino-cerebral aspergillosis. Orbital and cranial base aspergillosis can cause gradually progressive deterioration of vision. Angioinvasion in a case of invasive aspergillosis can lead to stroke like symptoms. The mainstay of treatment is surgical debridement with anti-fungal therapy. Although, Amphotericin-B is the traditional drug of choice but Voriconazole is better tolerated and has significantly less toxicity compared to Amphotericin-B.

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BRANCHIO-OTO-RENAL SYNDROME: A CASE REPORT & REVIEW OF LITERATURE

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ABSTRACT:

Background: Branchio-oto-renal (BOR) syndrome is a rare autosomal dominant disorder characterized by syndromic association of branchial arch anomalies, otologic malformations and renal anomalies. Very few articles on this syndrome have been published in journals related to Otorhinolaryngology.

Case Report: A 18 years old male patient, presented with a right sided intermittently discharging neck sinus at the junction of middle 1/3rd and lower 1/3rd of anterior border of sternocleidomastoid muscle along with bilateral pinna deformity and preauricular sinus. Fistulogram revealed a fistulous tract communicating with the right tonsillar bed. NCCT temporal bone showed marked hypoplasia of bilateral external auditory canal, dysplastic middle ear ossicles and microtia. NCCT KUB showed a hypoplastic right kidney and malrotated left kidney although his renal function test were within normal limits. In the presence of 3 major symptoms, branchial fistula, bilateral middle ear malformations and renal structure abnormality this case was clinically diagnosed as branchio-oto-renal syndrome. The patient was treated for his active symptoms hence his fistulous tract was excised after initial medical management.

Conclusion: The BOR syndrome is an infrequent but a well described entity that combines branchial arch anomalies, otologic malformations and renal anomalies. The BOR syndrome should be included in the differential diagnosis of deafness and chronic renal failure in childhood and adolescence.

Key words: Branchio-Oto-Renal Syndrome, fistula, branchial, renal malrotation, preauricular sinus, renal hypoplasia.

INTRODUCTION:

Branchio-oto-renal (BOR) syndrome is a rare autosomal dominant disorder with prevalence of approximately 1 in 40,000 new born infants and in about 2% of children with profound deafness^{1,2}. Branchio-oto-renal (BOR) syndrome was first described by Melnick et al³ in 1975 followed by Fraser et al⁴, thus giving the name Melnick-Fraser syndrome.

It is characterized by branchial arch anomalies (branchial clefts, fistula, cysts), hearing impairment (malformation of auricle with preauricular sinuses, conductive, sensorineural, or mixed hearing impairment), and renal malformations (urinary tract malformation, renal hypoplasia or agenesis, renal hypoplasia, renal cysts)⁵. Patients usually present with hearing impairment and discharging branchial fistula.

Such patients should be further investigated for BOR syndrome and thus control its progress.

Phenotypic presentation of BOR syndrome is extremely variable⁶. The combination of deafness with chronic renal failure may be confused with the Alport syndrome. Anterior lenticonus, a conical protrusion of the central portion of lens in to the anterior chamber is pathognomonic for Alport's syndrome. Moreover deafness in the Alport's syndrome manifests at a later age^{7,12}.

CASE REPORT:

An 18 years old male patient presented to the Otorhinolaryngology out-patient department with the chief complaints of right sided intermittently discharging neck sinus since birth.

He was born of non-consanguineous marriage and there was no history of hearing deficits, renal disease and similar discharging sinuses in the parents or in his only sibling. His birth history was insignificant.

On examination the patient was noted to have a discharging fistulous tract over right side of the neck at the junction of middle 1/3rd and lower 1/3rd on the anterior border of sternocleidomastoid muscle. The patient also had bilateral pinna deformity along with bilateral preauricular sinus.



Fig 1: showing pinna deformity and discharging sinus over right side of neck



Fig 2: Showing bilateral pinna deformity

Facial nerve examination was within normal limits on both sides and audiological assessment revealed bilateral severe mixed sensorineural hearing loss.

On investigating further, following were noted:
Fistulogram shows fluid contrast through fistulous tract communicating with the right tonsillar bed.

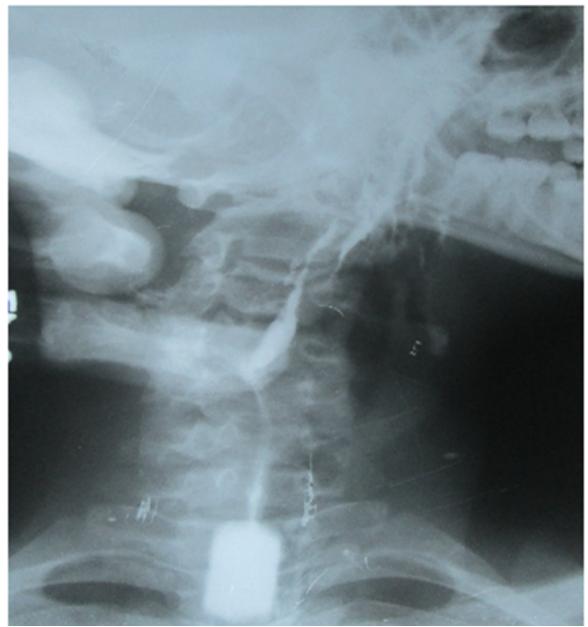


Fig 3: fistulogram

NCCT temporal bone shows marked hypoplasia of bilateral external auditory canal, dysplastic middle ear ossicles and bilateral microtia. Scans also revealed opacification of middle ear cavity on both sides.



Fig 4: NCCT Temporal Bone

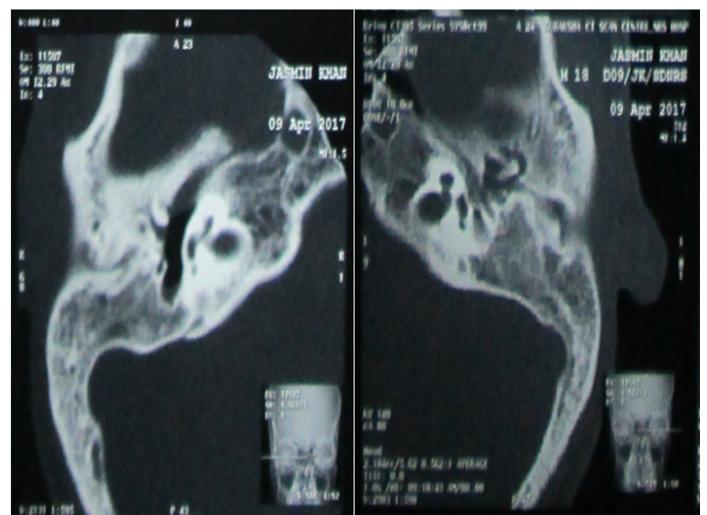


Fig 5 Right side

Although the patient had no complaints related to renal function, but on the basis of suspicion for BOR syndrome a NCCT KUB was requested and it revealed a small right kidney and malrotation of left kidney.

Fig 6 Left side

He was then referred to a nephrologist for his renal anomaly and subsequent regular follow up.

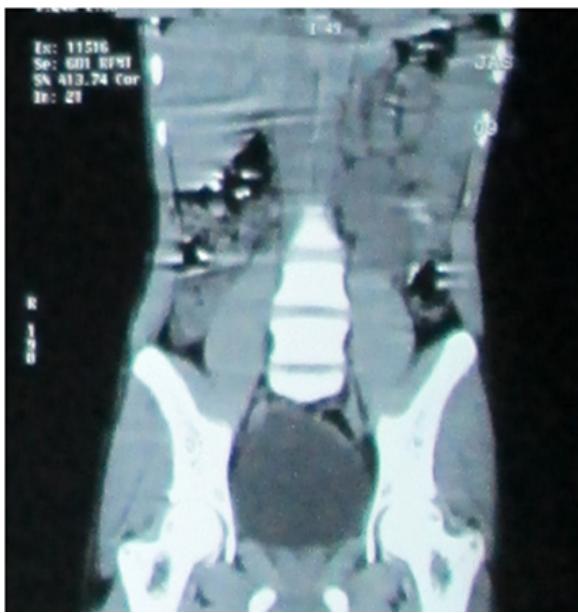


Fig 7 CT Abdomen

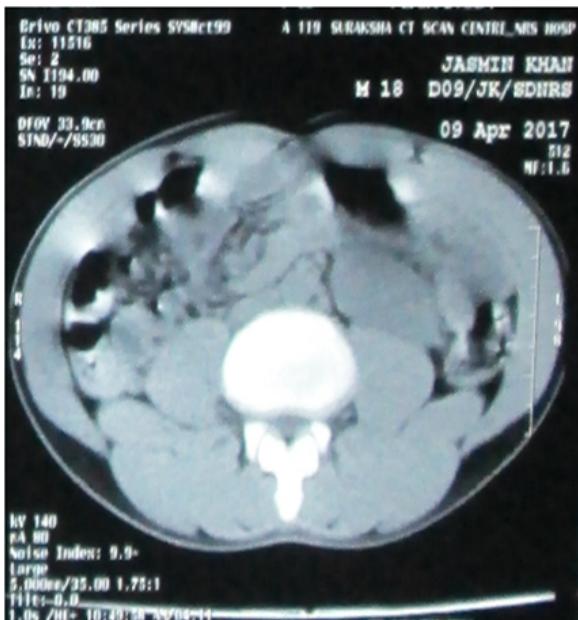


Fig 8: CT Abdomen

As a part of his management work up the patient was treated only for its active symptoms.

Hence the patient underwent complete excision of the fistulous tract after initial stabilization and medical management. He was advised hearing aids, regular hearing assessment, and periodic renal function monitoring.

In the light of 3 major symptoms, branchial fistula, bilateral middle ear malformations and renal structure abnormality this case was clinically diagnosed as Branchio-oto-renal syndrome.

DISCUSSION:

The BOR syndrome is a rare but well defined constellation of branchial arch anomalies (branchial fistulas, branchial cysts), external ear malformations with

hearing loss and renal hypoplasia and dysplasia. It has an autosomal dominant transmission pattern with variable clinical expression⁶

Chang et al. developed the diagnostic criteria of BOR syndrome in 2004^{8,9}. These include major criteria such as second branchial arch anomalies, hearing loss, preauricular pits, auricular deformity and renal anomalies, and minor criteria such as external auditory canal anomalies, middle ear anomalies, inner ear anomalies, preauricular tags and other symptoms such as facial asymmetry and palate abnormalities.

In patients with a family history, any single major criterion is sufficient for diagnosis of BOR syndrome. Without any family history, three major criteria or two major and two minor criteria are needed to make a confident diagnosis.

EYA1 (8q13.3), the human homologue of the *Drosophila* eyes absent gene, is the most frequent causative gene for BOR syndrome¹⁰. It expresses very early, between 4th & 6th weeks of human embryogenesis¹¹. Deafness relates to abnormalities in the three ossicles of the middle ear derived from the first and second branchial arches, while the branchial fistulae relates to second, third and fourth arches. EYA1 gene is strongly expressed in the human embryonic kidney and in BOR syndrome there is fault between the ureteric bud and metanephric mesenchymal mass as the ureteric bud branches into renal parenchyma, resulting in renal anomalies¹². Mutations in SIX1 (14q23.1) and SIX5 (19q13.32) have been reported less frequently¹³. SIX1 interacts with EYA1 in the development of various organs¹⁴. SIX5 has a high degree of homology to SIX1 and directly interacts with EYA1¹⁵.

Clinical features are highly variable. The most common presenting symptom is deafness (90%) which can be sensorineural or conductive but is mostly mixed (50%)¹⁶. Pre-auricular pits can be the presenting features in over 70% of the cases and sometimes can be the only external ear finding as seen in our patient, while around 50% of patients have external ear anomalies in the form of microtia to small lop or cupped ears with over folded superior helices¹⁷. Middle ear anomalies include ossicular malformations and inner ear anomalies include cochlear hypoplasia or dysplasia¹⁸. Renal anomalies include renal dysplasia and agenesis which can lead to end-stage renal disease¹⁹.

CONCLUSION:

BOR syndrome is a rare but well described clinical entity. It combines branchial anomalies with otologic and renal malformations. Most individuals with BOR syndrome do not have a life-threatening condition, and, in many families, it is not uncommon for the disease to go undiagnosed until the birth of a child with severe

manifestations of the BOR phenotype. This is unfortunate, as recognition of the hallmark features of BOR syndrome could ensure that affected persons receive appropriate medical information and care. Integral elements of medical care include audiologic, otologic, head & neck, urologic, and genetic evaluation. Patients with BOR syndrome who receive adequate treatment can lead normal, productive lives. It is therefore essential to provide early diagnosis including the relevant genetic tests, although further studies are needed to unearth the molecular mechanisms and undiscovered causative genes responsible for BOR syndrome.

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