Endolymphatic Sac Tumor in VHL Management and Audiological Rehabilitation

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Endolymphatic sac tumors (ELST) are rare, locally aggressive papillary tumors, histologically benign, growing from inner ear. Although they can present as sporadic tumors, this particular neoplasia is more usually diagnosed in VHL patients (10-15 %). In VHL patients, bilateral ELST is a unique finding, in 11% of affected cases (Poulsen et al).

ELS tumors produce hearing impairment and vestibular disfunction (Menière like) through otic capsule injury, endolymphatic hydrops and/or intralabyrinthine hemorrhage .

Classical management of this tumor has been wait and see up to neurological symptoms, because its indolent growth, but considering hearing loss an unavoidable consequence.

More recently, image and surgical improvements have shown that severe hypoacusia can be avoided with early diagnosis and tumor resection.

Therefore, after Megerian´s (2002) description of surgical strategies, Lonser et al have changed and reversed the paradigm of treatment, advicing early diagnosis and surgery when at least partial hearing can be preserved.

But many patients are still diagnosed when hearing is severely impaired, or when tumors are big enough that resection will be followed by deafness.

OBJECTIVE:

Review the present ELS tumor management and audiological rehabilitation maneuvers in VHL patients diagnosed and treated in a multidisciplinary VHL unit.

JMG. 43y old female. Mother and three other siblings affected.

Symptoms started when 12y old, with successive multiple Hemangioblastomas in retina (rigth blindness, left restriction) and in cerebellum. Pancreatic and parametrial cysts. Multinodular thyroid hyperplasia.

Progressive right hypoacusia, tinnitus, vertigo and disequilibrium in the last 10 years.

2006.- Right ELST, resected through mastoidectomy, with little improvement. Left progressive hypoacusia.

2009.- FJD.

Audiological studies:

Rigth: Hypoacusia 90 dB, cochlear reserve 25 dB

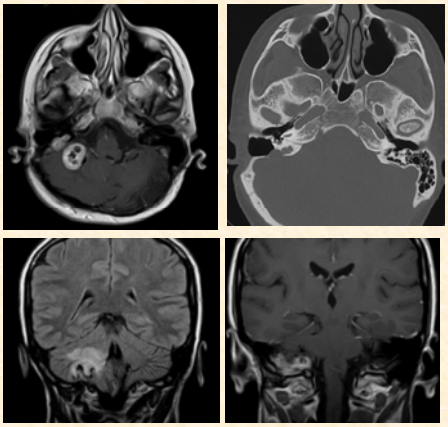
Left: Hypoacusia 25 dB, cochlear reserve 25 dB

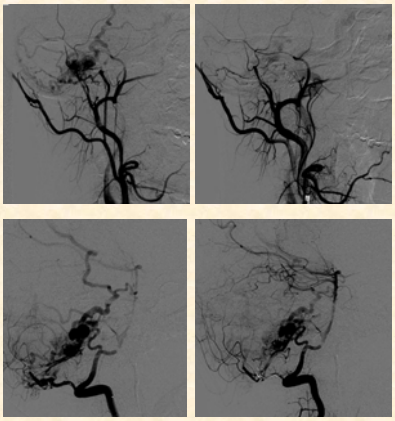
Tinnitus perception on right ear at 3000 Hz with 45 dB

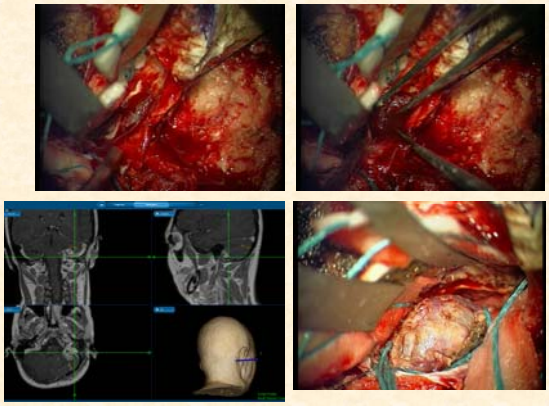
Conclusions:

Severe Right Hypoacusia, with preservation of bone way speech comprehension

Mild Left Neurosensorial Hypoacusia.







TREATMENT:

Tumor resections through a combined retrosigmoid approach and reopening of mastoidectomy.

One year later, without tinnitus and vertigo, and no tumor in image study:

Right Osteointegrated (**B.A.H.A**) implant.

Binaural hearing sensation and speech discrimination improved

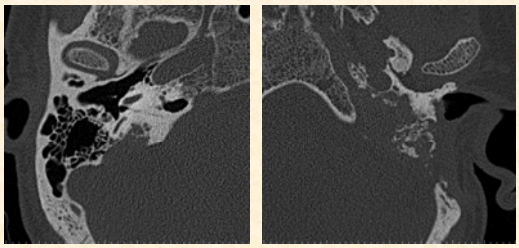
FMS: 41y old male. No other VHL affected members in the familiy.

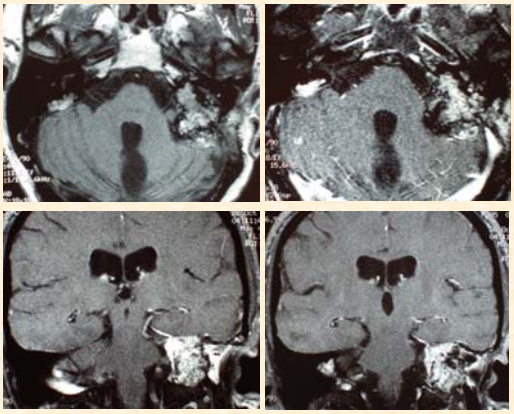
Since the age of 13, he has been treated for cerebellar hemangioblastomas (3 resections, disequilibrium), bilateral CCRC (right nefrectomy, selective tumorectomies, left nefrectomy, chronic hemodyalisis since 2004), left retinal hemangioblastoma.

Since the age of 24, progressive bilateral hypoacusia, tinnitus and aural fullness. In 2004, a huge left petrosal tumor was diagnosed and partially resected, stopping surgical procedure because hemorrhage. At biopsy, “papillar adenocarcinoma”.

2007.- Endovascular embolization and radiotherapy. Binaural hearing loss and unbearable tinnitus.

2010.- FJD





TREATMENT:

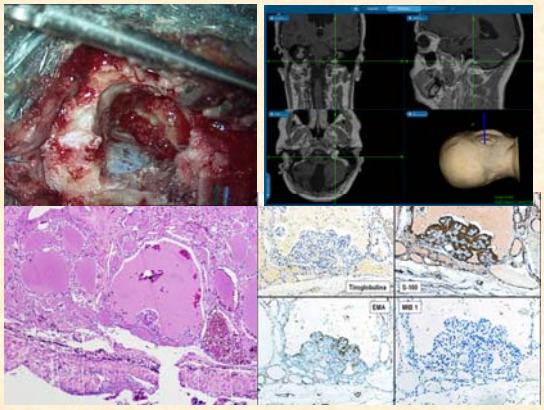
Endovascular embolization of the right tumor (branches of posterior auricular artery).

Surgical Approach through Right Retrolabyrinthine Posterior Petrosectomy, resecting tumor and adjacent presigmoid duramater, and placing a Nucleus 24 cochlear implant, with magnet removed (external adhesion).

Audiological rehabilitation: relevant improvement, with 75% speech discrimination , and 40 dB in audiometry.

Left tumor volume reduced . Tinnitus reduced.

Awaiting renal transplantation.



Audiological invasive aids for severe hearing loss after ELS tumor treatment:

Cochlear Implantation for Hearing Loss Associated With Bilateral Endolymphatic Sac Tumors in von Hippel-Lindau Disease

*†Jay Jagannathan, *Russell R. Lonser, *Richard A. Stanger, ‡John A. Butman, *Alexander O. Vortmeyer, §Christopher K. Zalewski, §Carmen Brewer, ‖Christal Surowicz, and §¶H. Jeffrey Kim

Cochlear implantation in a bilateral endolymphatic sac tumor patient
A case report

Carlos M. Boccio, Gabriela M. Pérez Raffo *, Cecilia Parsini

Cochlear Implantation After Transmastoid Labyrinthectomy

John F. Kveton, MD; Cynthia Abbott, MEd; Max April, MD; Geralyn Drumheller, MEd; Nancy Cohen MEd; Dennis S. Poe, MD

CONCLUSIONS:

ELS tumors should be resected early, when diagnosed.

Presurgical endovascular embolization of extracranial arterial afferents can make surgery easier.

Resection of tumor with resection of neighbor posterior fossa dura avoids recurrences.

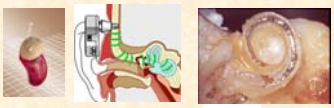
CONCLUSIONS:

THINK about posible audiological rehabilitation after tumor surgery, improving the Q o L.

Unilateral mild hypoacusia: External hearing aid.

Unilateral severe hypoacusia: Osteointegrated hearing aid, best if cochlear function remnant.

Bilateral severe hypoacusia, with cochlear activity: Cochlear implant.



Thank you !!!




SEE YOU IN VHL MADRID 2014!!!!