A Case Report of Skull Base Chondrosarcoma: An Appraisal

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Abstract

Background: *A rare slow-growing skull base tumor with poor outcome due to compression of adjacent structures is chondrosarcoma.*

Materials and Methods: *We describe one case of chondrosarcoma who initially presented to us with features of trigeminal neuralgia. Magnetic resonance imaging (MRI) revealed a skull base tumor. Though total surgical excision is the aim, subtotal resection was only possible in our case due to local anatomical limitations and involvement of important neurovascular structures.*

Results: *The histo-pathologic examination revealed chondrosarcoma.*

Conclusion: The differential diagnosis for chondrosarcoma in the skull base region is chordoma and chondroid chordoma which have different prognoses. In conclusion, accurate diagnosis and careful surgical treatment play important roles in the further management of chondrosarcoma.

Key Words: *chondrosarcoma; chordroma; Skull base tumor.*

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Introduction:

Chondrosarcoma of the skull base is a slow-growing indolent tumor with a potentially lethal outcome due to compression of adjacent tissues such as the carotid artery and cranial nerves. Radical excision is usually difficult. It accounts for 6% of all skull base lesions[1].Chondrosarcoma is mostly divided into three histological grades, grade I (well differentiated), grade II (moderately differentiated), and grade III (poorly differentiated). The 5-year survival rates for grade I, II and III chondrosarcomas of bone from all body sites are 90%, 81%, and 43%, respectively[2]. Surgical treatment with adjuvant radiotherapy, particularly carbon ion radiotherapy, has been reported to achieve a better outcome than simple excision of tumor[3].

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Case Report:

A 48 years old man presented to us with complaints of episodic severe pain in the right eye, upper half of the face, diplopia for the last 2 months and decreased hearing on right for last 1 month. His past history was not significant. On examination, his right pupil was dilated, fixed and not reacting to light, complete opthalmoplegia, complete ptosis and proptosis of right eye. He had decreased sensation in all the three divisions of CN III in the left eye.

MRI brain with contrast showed heterogeneously enhancing lesion involving the clivus, right subtemporal area, sellar, para-sellar and supra-sellar region. It extended to the retro-orbital area and pushing internal carotid artery superomedially (Figure 1). Tumor was removed sub-totally leaving cavernous and carotid portion of the tumor behind. Tumor was moderately vascular and firm in consistency.

Histopathology shows neoplasm, composed of cartilaginous tissue, spicules of bone. Focal areas of increased cellularity was present with slight pleomorphism, which was consistent with low grade chondrosarcoma (Figure 2). Further evaluation with immuno-histochemical staining was not performed.

In the post-operative period, he was free from right orbital pain but had developed chemosis of the right eye which subsided over one week. Patient was given radiotherapy post-operatively. After receiving radiotherapy, his ptosis improved and he continues to visit us an outpatient every 3 months for follow-up.

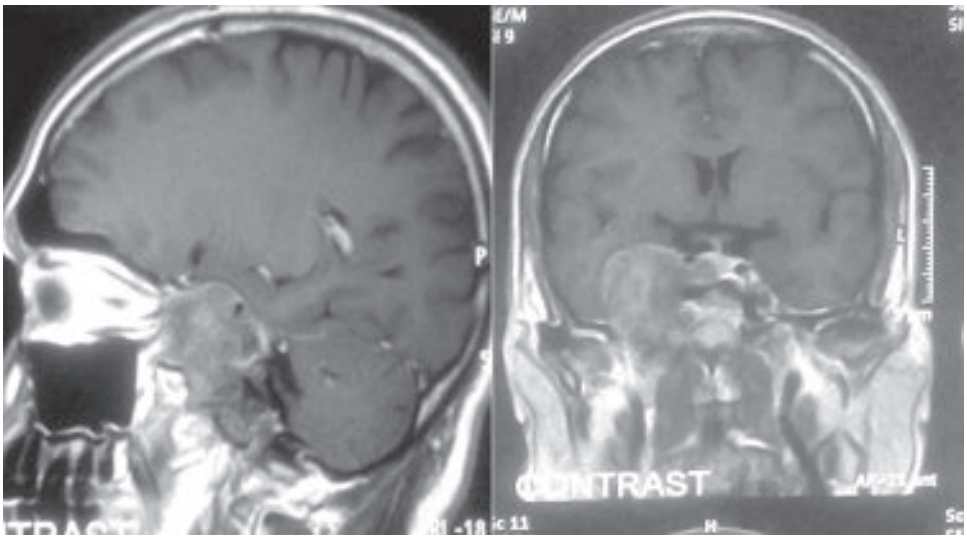

Fig.-1: *MRIbrain with gadolinium contrast administration (Sagittal and coronalkey images) shows heterogeneously contrast enhancing lesion involving clivus , right subtemporal area ,sellar, para-sellar and supra-sellar; with extension to the retro- orbital region and pushing internal carotid artery superomedially.*

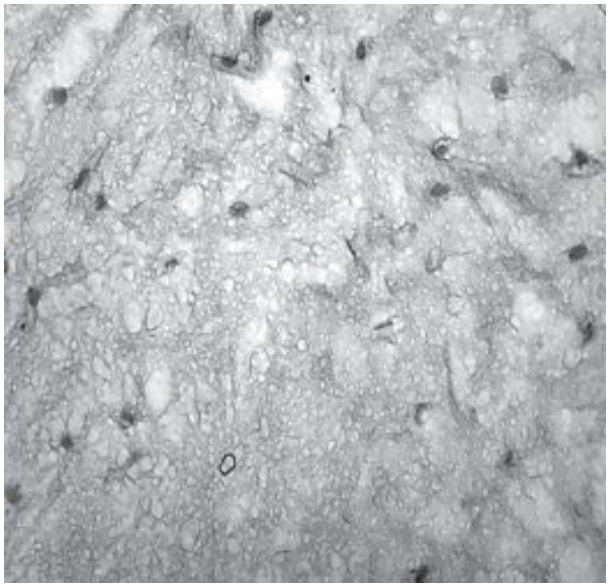

Fig.-2: *Histopathology (Haematoxylin &Eosin 40X magnification) is consistent with Low grade chondrosarcoma .Background consisted of cartilaginous matrix with focal areasof increased cellularity andcellular pleomorphism.*

Discussion:

Chondrosarcoma of the skull base is rare and accounts for 6% of all skull base lesions[1] and 0.1% of all head and neck tumors [4]. Chondrosarcoma arises from the remnant of fetal cartilage and notochord in the skull base or from pluripotent mesenchymal cells involved in the embryonogenesis of the skull base.

Chondrosarcoma can be further sub classified, in order of frequency, into the conventional (hyaline or myxoid), dedifferentiated, clear cell, and mesenchymal subtypes. Hyaline chondrosarcoma is the most common subtype[5]. Dedifferentiated chondrosarcoma is the most malignant variant with a high risk of metastasis. It is difficult to differentiate chondrosarcoma of the skull base from chordoma radiologically and histopathologially. Although they are similar in management, distinction between chordoma and chondrosarcoma is important due to different prognoses and outcomes[6]. A chordoma typically contains cohesive nests and cords of large cells with bubbly eosinophilic cytoplasm called physaliphorous cells. Although physaliphorous cells may be present in chondrosarcoma, they are smaller and have less cytoplasm than those seen in a chordoma, and lack cohesive nests or cords. Chondroid chordoma has features of both chondrosarcoma and chordoma. Chondroid chordoma is similar to chondrosarcoma in the cartilaginous areas and contain the cohesive nests and cords of physaliphorous cells that are typical features of chordoma [7].

Immunohistochemical study is helpful to differentiate chondrosarcoma from chordoma .Chondrosarcoma is usually positive for S-100 and negative for EMA and cytokeratin (CK) whereas Chordoma, is usually positive for EMA, CK and S-100[8]. Diffuse chordoma

background with areas of chondroid patterns immunonegative for CK staining is more suggestive of chondroid chordoma[9].

In our case, we did not perform immunohistochemical staining for S-100, EMA and CK which could have further bolstered the diagnosis of chondrosarcoma which was our pathological diagnosis. Immunohistochemical staining is undoubdetly of importance as chordoma and chondrosarcoma can have similar view under the microscope and histopathologist may have hard time differentiating the two. Hence, the learning curve is that it is empirical we perform further inmmunohistochemical analysis to bolster/re-confirm the histological diagnosis as the prognosis and treatment of chondrosarcoma and chondroma are different. The former has more malignant behaviour.

Treatment of chondrosarcoma includes careful preoperative evaluation and surgical resection with adjuvant radiotherapy which has been reported to achieve a better outcome than simple local control by surgical resection[3].Computed tomography (CT) and MRI are helpful tools to demonstrate bony involvement, calcification in the rim of the lesion and soft tissue delineation respectively which are important for surgical planning[10].

Conclusion:

A chondrosarcoma of the skull base is a rare, slowly growing tumor which should always be differentiated from chordoma due to different clinical outcomes. It is imperative that a pathological diagnosis of either two different tumours being re-confirmed with further immunohistochemical analysis. This is of paramount importance as the prognosis of these two different tumours varies and patient can be counselled and treated accordingly based on the correct histological diagnosis. Local control by radical excision followed by radiotherapy is the current recommended management treatment protocol for chondrosarcoma.

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