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Low-Grade Chondrosarcoma Mimicking Chondroid Chordoma: A Case Report

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ABSTRACT

Chondrosarcomas and Chordomas account for the majority of primary skull base tumors, which are slow growing and locally aggressive. These tumors cause severe cranial nerve neuropathies. Both these tumors have similar clinical, radiological, and histological resemblances. Histopathology, along with immunohistochemistry, helps to differentiate these tumors. We are presenting a case of 39year old female who came with double vision, squint, headache, and tingling sensation over the right side of the face. Examination and investigations concluded a skull base tumor near the cavernous sinus. Surgical resection of the lesion was done and sent for histopathology. Microscopy showed hyaline cartilaginous islands, myxoid matrix, and individual cells with vacuolated cytoplasm and ovoid nucleus. Immunohistochemistry showed positive for S100. Final diagnosis of low-grade Chondrosarcoma was made.

Keywords: Chondrosarcoma, Skull base tumors, Chordoma, S100

INTRODUCTION

Intracranial Chondrosarcomas are very rare, accounting for 0.15% of all intracranial neoplasms [(1)]. The prevalence of Chondrosarcomas arising from the skull base is only 0.1-0.2% which constitutes 2% of all Chondrosarcoma cases [(2,3)]. Both Chondrosarcomas and Chordomas have similar characteristics on CT and MRI, making them difficult to differentiate from one another [(4)]. Majority of the Chondrosarcomas of the skull base are located away from the midline, which is a helpful sign to differentiate them from chordomas, which are mostly midline. Most of the time, the clinical presentation is due to the symptoms related to direct compression of cranial nerves. The surgery mainly helps in the histological diagnosis and also relieves the pressure symptoms. Adjuvant radiotherapy is often administered to treat residual disease and prevent recurrence.

Here we present a case of low grade Chondrosarcoma, which mimics Chondroid chordoma in histology.

CASE HISTORY

A 39year old female came with complaints of double vision, headache, squint, and tingling sensation on the right side of face for one year. On examination, the Glasgow coma scale shows E4V5M6. Power is 5/5 in all limbs. Perception of light is present. Pupils are reactive to light. Eye movement is restricted on the right side. Right sixth cranial nerve palsy is noted on examination. MRI showed a 3.5x2.8cm large enhancing left cavernous sinus lesion with extension to superior orbital fissure and encasing optic nerve. Clinical diagnosis of CP angle tumor was made. Right temporal craniotomy is done. Intraoperative findings showed a greyish-white soft lesion showing partial calcifications. Near total excision was done

and sent for histopathology. In the pathology department, we received multiple grey-white soft tissue bits measuring 5x5cm. Microscopy shows tumor tissue arranged in a lobular and nesting pattern, separated by fibrous strands, tumor cells having round to oval nuclei with abundant clear cytoplasm, with some of them showing vacuolated appearance and separated by myxoid matrix. At some foci, the matrix shows hyaline cartilage with areas of calcification. Histologically differential diagnoses of low-grade Chondrosarcoma and Chondroid chordoma are considered. Immunohistochemistry results showed tumor cells positive for S-100 and negative for EMA. Final diagnosis of low grade chondrosarcoma was made.

DISCUSSION

Chondrosarcomas are malignant tumors that are characterized by the production of cartilaginous matrix. The pathogenesis of skull-base Chondrosarcomas is linked to the endochondral ossification of the skull-base synchondroses[5]. Clivus and temporo-occipital junction are the most common sites. In most cases, Chondrosarcomas affect the clivus and spread to the middle cranial fossa around the sellar region (30-50%) and the posterior cranial fossa (50%) [6]. Histologically conventional Chondrosarcomas range from WHO grade I (well differentiated) to WHO II and III (poorly differentiated) tumors[7,8]. Microscopically Classical conventional grade I Chondrosarcoma shows chondrocytes embedded in a cartilaginous matrix with minimally increased cellularity, nodular growth, and occasional binucleate cells. Grade II tumors show moderate cellularity and diffuse growth. Grade III tumors show highly increased cellularity, many atypical cells, pleomorphic appearance, and easily identifiable mitotic figures. Grade I are the most common cranial Chondrosarcoma accounting for 80% of cases [9].

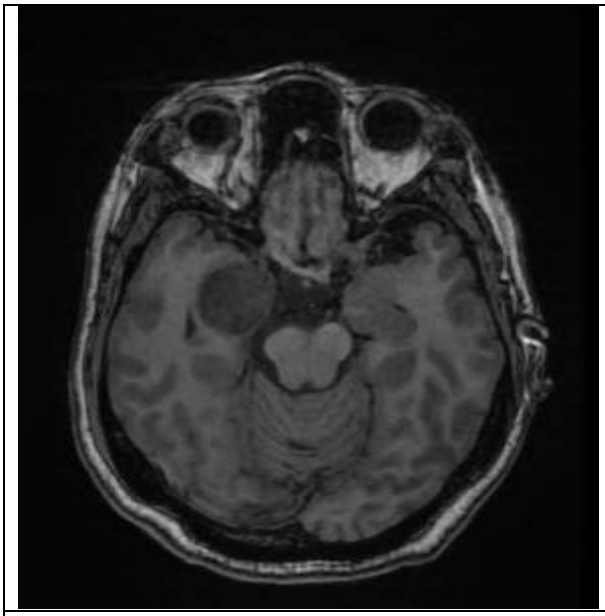

Figure 1: MRI brain showing 3.5X2.8cm large enhancing cavernous sinus lesion extending to superior orbital fissure and encasing optic nerve

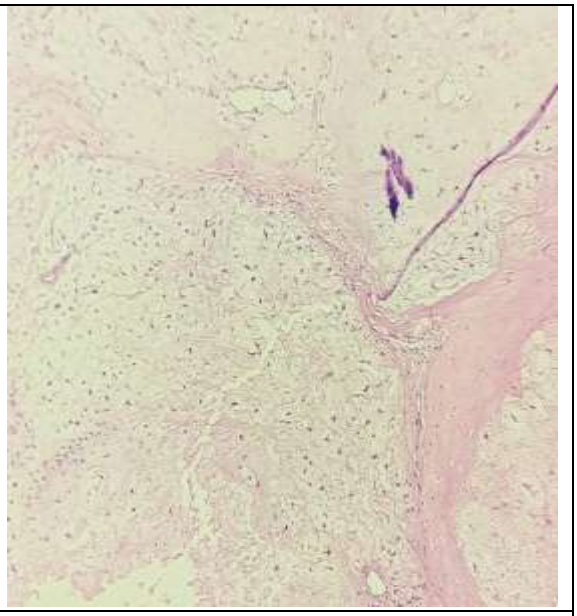

Figure 2: Microscopy shows tumors cells arranged in lobules and nests separated by fibrous septa (X100; H&E)

Chordomas are locally aggressive neoplasms arising from the notochordal remnants. The first macro-microscopic description of Chordomas was provided by the German physician and pathologist Rudolf Virchow [10]. The histology of these tumors has unique cells, called physaliferous cells, which are characterized by intracellular, bubble-like vacuoles. There are three microscopic subtypes of chordoma based on morphology – conventional, chondroid, and dedifferentiated chordomas.

Out of these three, Chondroid Chordoma is associated with favorable survival rates, and dedifferentiated Chordomas have an unfavorable survival [(11)]. Histologically Chondroid Chordoma shows areas of chondroid differentiation along with conventional physaliferous cells and myxoid matrix.

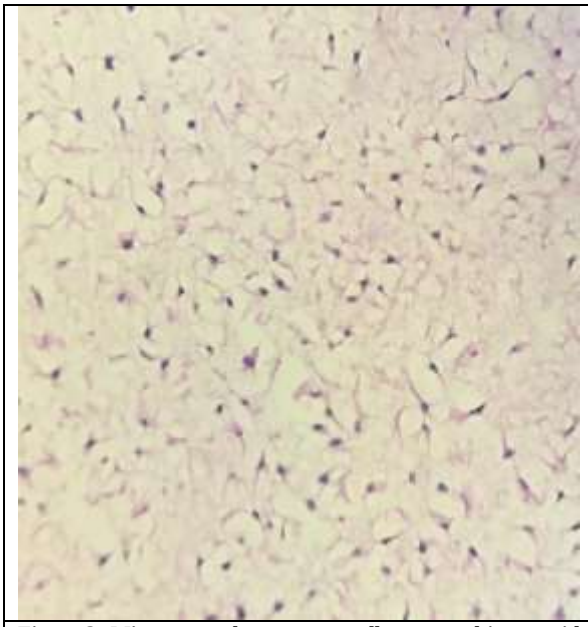

Figure 3: Microscopy shows tumor cells arranged in myxoid matrix (X400; H&E)

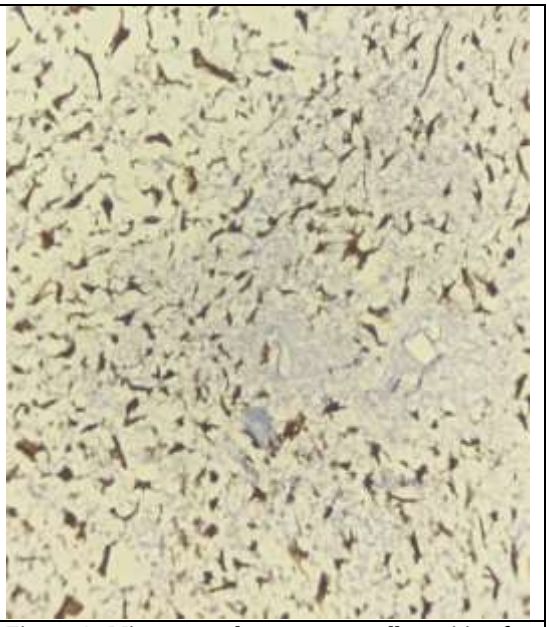

Figure 4: Microscopy shows tumors cells positive for S100 immunohistochemistry (X100; H&E)

High and specific immunohistochemical expression of brachyury along with positivity for EMA is seen in Chordomas, and it helps in distinguishing Chordomas from Chondrosarcoma. Chondrosarcoma shows positivity to S 100 immunostaining.

CONCLUSION

Skull base tumors are very rare and aggressive tumors, where surgical management of the patient is very difficult. Appropriate radiological investigations and histopathological examination are very important to provide any adjuvant therapy to the patient. Use of basic immunohistochemistry markers like S100 and EMA aids in the final diagnosis whenever there is a diagnostic dilemma between Chondrosarcoma and Chordoma. We present this case because of its diagnostic difficulty and resemblance to chordoma.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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