International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Chondrosarcoma of Jugular Foramen- A Rare Case Report

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Received: 06/01/2015 *Revised: 27/01/2015* *Accepted: 27/01/2015*

ABSTRACT

Skull base tumors are rare and challenging for diagnosis and treatment. Chondrosarcoma is a rare malignant neoplasm of skull base. We present a case of 21 year old male having chondrosarcoma of skull base arising from jugular foramen, eroding adjacent bony structures and reaching lower level of C1 vertebra. It needs to be differentiated from chordoma due to its same anatomic location. This has important clinical implications, because when treated with similar aggressive treatment strategies, chondrosarcomas have better prognosis than chordoma. In our case, immunohistochemical study of the tumor was positive for S100 protein and negative for cytokeratin and epithelial membrane antigen which helps in differentiating from other lesions. Herein, we describe a case of classic chondrosarcoma grade I of skull bone for its clinical, radiological, histopathological and immunohistochemical study.

Keywords: foramen jugular, chondrosarcoma, immunohistochemistry.

INTRODUCTION

Chondrosarcoma is a malignant tumor with pure hyaline cartilage differentiation. Chondrosarcoma accounts for 6% of skull base neoplasms and 0.15% of all intracranial tumors. [1] These are slow growing indolent tumors with potentially lethal outcome due to compression of adjacent tissues such as carotid artery and cranial nerves.

CASE REPORT

A 21-year-old male patient complaining of neck pain, hoarseness of voice and irritability of 6 month duration. Computed tomography (CT scan) showed large hypodense defect involving right jugular foramen causing with multiple foci. Punctate curvilinear areas of calcification extension from lower C1 through jugular foramen up to sigmoid sinus causing smooth expansion of jugular foramen.(figure1) Patient was subjected to MRI(plain and contrast) which showed large well defined multilobulated extra axial mass lesion extending from right jugular foramen superiorly reaching up to right cerebello-pontine angle and tentorium cerebelli causing adjacent mass effect and erosion of adjacent bony structures followed by contrast. A erosion of petrous part of right temporal lobe and scalloping of the clivus

laterally on right, involving the occipital bone causing periosteal reaction. Other routine investigations were in normal limit and there was no significant contributory history. The tumor mass was removed surgically and send for histopathological examinations.

Gross examination: We received multiple, irregular, grey-white, firm to hard tissue pieces aggregating to 5.2x3.1x2cm and the larger one measured 2x1cm. Cut surface showed grey-white, hard, pearly white areas.

Microscopic examination: The sections showed bone and soft tissue with an ill-defined malignant cartilaginous neoplasm arranged in lobular pattern with features of low grade classic chondrosarcoma (figure 2). Tumor showed proliferation of chondrocytes containing hyperchromatic nuclei and moderate amount of eosinophilic cytoplasm (figure 3). Occasional binucleate cells were seen. Tumor invasion into bone and intra-trabecular areas were noted. There was no definite evidence of granulomatous inflammation. On immunohistochemistry neoplastic cells expressed S100 and they were negative for CK, EMA. The Mib-1 (ki 67) labelling index was low. The post operative outcome of the patient is well and is on regular follow up.

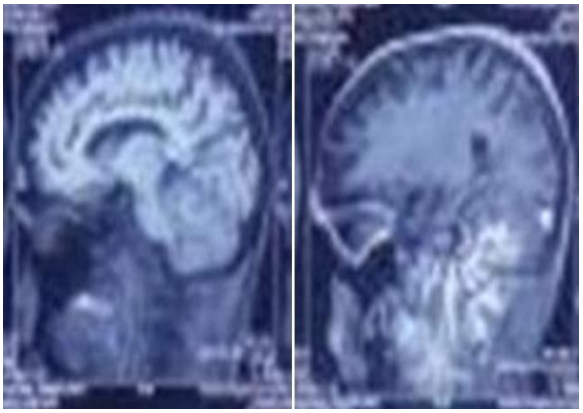

Fig 1: MRI showed large well defined multilobulated extra axial mass lesion extending from right jugular foramen.

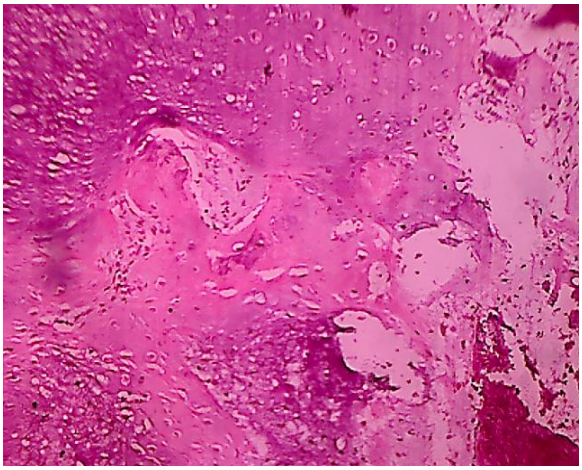

Fig 2: Photomicrograph showing bone and soft tissue with an ill-defined cartilaginous neoplasm arranged in lobular pattern with features of low grade classic chondrosarcoma. (H&E Stain, 40x)

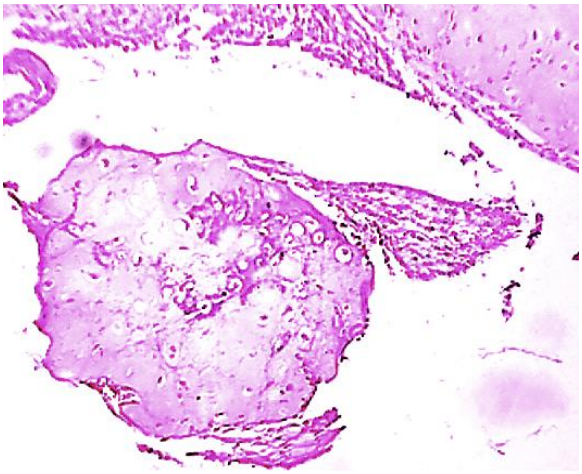

Fig 3: Photomicrograph showing proliferation of chondrocytes containing hyperchromatic nuclei and moderate amount of eosinophilic cytoplasm. (H&E Stain, 100x)

DISCUSSION

Primary bone tumors are classified on the basis of the appearance of the tumor cell, its product, or both. Chondrogenic tumors include osteochondroma, enchondroma, chondroblastoma, chondromyxoid fibroma and chondrosarcoma. [2]

Several hypotheses have been elicited to explain how a chondrosarcoma develops in the skull base. The histogensis of chondrosarcoma is postulated either to associate with the remnant of the fetal cartilage and notochord in the skull base or to arise from pluripotent mesenchymal cells

involving the embryogenesis of the skull base. [3] Chondrosarcoma can be subclassified, in order of frequency, into the conventional (hyaline or myxoid), differentiated, clear cell and mesenchymal subtypes. According to Rosenberg et al, conventional chondrosarcoma is the most common subtype. [4]

Classic chondrosarcomas are divided into grades I, II, or III based on mitotic rates, cellularity, nuclear size of the cells, and chondroid matrix. [5] Grade I: low-grade chondrosarcomas are cytologically so similar to enchondromas that the diagnosis is mostly dependent on the clinical and radiologic presentation, and on the location. Grade II chondrosarcoma shows a definite increased cellularity with increased nuclear size and distinct nucleoli in many of the cells. Binucleate cells are common. Focal myxoid chondrosarcomas are generally assigned grade II. Grade III: high-grade chondrosarcomas are comparatively rare. They are generally rapidly growing, aggressive, and frequently metastasis. In our case it is of classic type with low grade cellular features. Intracranial chondrosarcomas are typically of classical type and are low grade I (51%) followed by the grade II(11%) tumors followed by mesenchymal type(30%). [4,6]

We have to differentiate other condition on histopathological examination. The most important differential diagnosis for chondrosarcoma of the skull base is chordoma. Although they are similar in management, distinction between chordoma and chondrosarcoma is important due to different prognosis and outcome. 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. Chondroid chordoma is similar to chondrosarcoma of in cartilaginous areas and contains nests of cohesive cells or cords of physalifarous cells that are typical of chordoma. [7] In our case sections were devoid of physaliphorous cells.

Immunohistochemical study is helpful in puzzling cases. Chondrosarcoma is usually positive for S-100 and negative for epithelial membrane antigen (EMA) and cytokeratin (CK). [3] Chordoma, in contrast, is usually positive for EMA, CK. On immunohistochemistry, our sections were positive for S100 and negative for CK, EMA which is exactly same as studied.

Treatment of chondrosarcoma includes careful preoperative evaluation and surgical resection or radiotherapy, particularly carbon ion radiotherapy which has been reported to achieve a better outcome than simple local control. [8]

CONCLUSION

Chondrosarcoma of skull base is a rare, slowly growing malignant tumor. It should be differentiated by from chordoma due to different clinical outcomes by proper histological diagnosis followed by immunohistochemical studies. It should be promptly imaged preoperatively with the help of imaging modalities treated by effective surgery and if required radiotherapy.

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How to cite this article: Chougule MV, Pawar V, Jagtap SV et. al. Chondrosarcoma of jugular foramen- a rare case report. Int J Health Sci Res. 2015; 5(2):475-478.

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