

Case Report

An unusual case of extensive chondrosarcoma of nasal septum

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ABSTRACT

Chondrosarcoma is a very rare malignant tumour of nasal cavity. It is seen commonly in middle aged men. The definitive treatment is surgical resection with wide margins. Radiation and chemotherapy are reserved for residual or recurrent disease and as palliative treatment. Here we present a case of Grade II mesenchymal chondrosarcoma involving both nasal cavities extending into ethmoid, frontal and sphenoid sinuses with intracranial extension and also its surgical management.

Keywords: Chondrosarcoma, Malignant tumour, Surgery, Metastasis

INTRODUCTION

Chondrosarcoma is a malignant tumour of mesenchymal origin. It mostly affects pelvis, ribs and long bones. The head and neck region are rarely affected.¹ Chondrosarcomas are histologically similar to chondromyxoid fibromas and chondroid chordomas. They must be identified properly as the management differs and so is the long-term prognosis.² Chondrosarcomas of head and neck are associated with better disease-specific survival and overall survival rates.³

CASE REPORT

A 24-year old male presented to our outpatient unit with 6 months history of proptosis and 4 months history of progressive nasal obstruction. Diagnostic nasal endoscopy showed smooth pinkish mass lesion completely filling both nasal cavities (Figure 1). Further examination showed restricted movement of left eyeball and reduced visual field. Routine blood investigations turned out to be normal.

MR imaging was done which showed a large T2 weighted hyperdense mass lesion in the midline involving ethmoid and frontal sinuses extending into the

left frontal lobe with break in the cribriform plate without any brain oedema. The lesion was hypodense in T1 weighted image and having strong non-homogenous T1 weighted post contrast (gadolinium) enhancement. Posteriorly the lesion extends into sphenoid sinus and inferiorly to the nasopharynx (Figure 2) There is no intradural involvement. The tumour compresses the left globe causing proptosis, but the carotids on both sides are spared (Figure 3).



Figure 1: (A) clinical examination showing proptosis, (B) nasal endoscopy picture showing tumour.

A computed tomography of paranasal sinuses and orbit was taken which showed destruction of floor of sphenoid bone, bilateral ethmoid trabeculae, perpendicular plate of

ethmoid, roof and medial wall of left orbit. There were small focal areas of calcifications within the lesion at places with destruction of nasal septum.

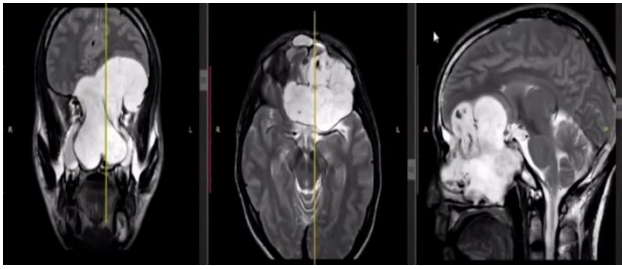


Figure 2: MRI imaging showing the tumour extend.



Figure 3: (A) Extradural location of tumour (B) tumour compressing the globe (C) the tumour sparing the carotid arteries on both sides.

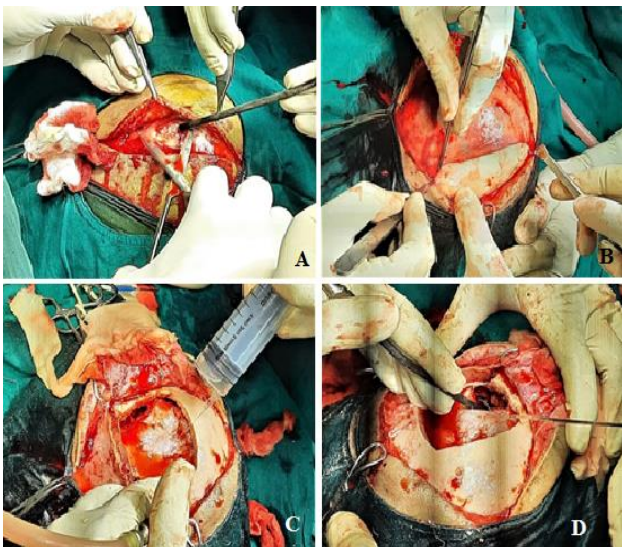


Figure 4: (A) Bicoronal incision (B) elevation of pericranial flap (C) bone window created (D) tumour removal and pericranial flap reconstruction.

A biopsy was done which showed spindle cells admixed with lobular grey blue abundant chondroid matrix, with atypical chondrocytes which were varying in size, shape, containing enlarged hyperchromatic and moderately pleomorphic nuclei and frequent binucleation, suggestive of a grade II chondrosarcoma.

So, we planned complete surgical resection of tumour with an aim to minimize any neurologic sequelae. A

combined approach, primarily the endonasal endoscopic debulking of tumour with powered instruments and also with a bicoronal frontal craniotomy approach to remove the remaining tumour and involved dura along with a pericranial flap reconstruction were done (Figure 4). Dural defect was closed with temporalis fascia. Intraoperative pictures showing tumour removal (Figure 5).

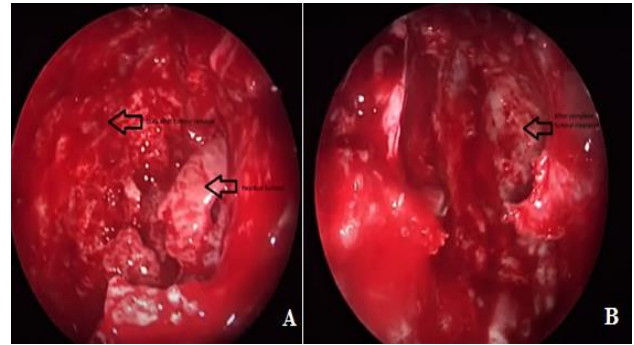


Figure 5: (A) Partial tumour removal (B) complete tumour removal.

Nasal packing was done. Negative suction drain was fixed and the bicoronal incision was closed in layers and dressing was applied. A lumbar drain was inserted to reduce the cerebro-spinal fluid (CSF) pressure. Foley's catheterization was done. Patient was put on triple antibiotic cover, anti-inflammatory agents, anti-epileptic drug and acetazolamide to reduce the CSF pressure. Patient was on strict bed rest in post-operative intensive care unit (ICU) for 5 days after surgery and he was given stool softeners and cough suppressants. The dressing was changed daily. On the 5th post-operative day, nasal pack was removed, nasal cleaning was done. The suction drain, the lumbar drain and Foley's catheter were removed. He was observed for one more day in the ICU. Patient was discharged on post-op day 7 in a stable condition without any CSF leak or infection.

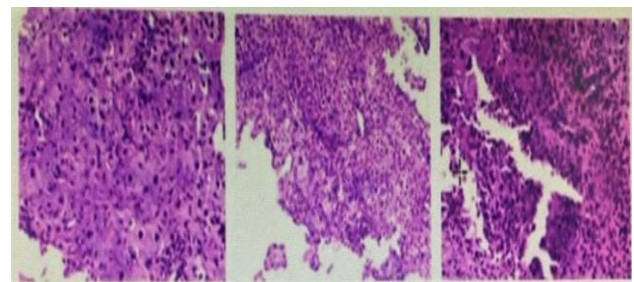


Figure 6: Spindle cells with lobular grey blue abundant chondroid matrix are seen. Atypical chondrocytes with pleomorphic nuclei are present.

The final histopathology report came as grade II mesenchymal chondrosarcoma (Figure 6). The patient was advised to undergo radiation therapy and was referred to a specialist. Post-surgery one-month evaluation was uneventful.

DISCUSSION

Chondrosarcomas in general are slow growing, locally aggressive tumours. These are very rare, constituting 10-20% of all primary bone tumours. Most common sites include pelvis, long bones and scapula. Head and neck chondrosarcomas are even rarer. They constitute only 4% on non-epithelial tumours of nasal cavity, paranasal sinuses and nasopharynx.^{1,4,5} Maxilla and mandible are the commonest sites in head and neck region. 0.05-41% of the head and neck chondrosarcomas originate from nasal septum.⁶

Chondrosarcoma is commonly seen among males with male to female ratio ranging from 1:1 to 10:1.^{1,6} Patients usually present with nasal symptoms such as nasal obstruction, epistaxis, rhinorrhoea, anosmia, head ache and ocular symptoms like exophthalmos, diplopia or epiphora.⁶⁻⁸ Chondrosarcoma arise in tissue formed of cartilage from rests of cells that remain after ossification, explaining the possible origin in nasal septum at the junction of sphenoid with perpendicular plate of ethmoid and vomer.^{3,9}

According to Lichtenstein and Jaffe, this tumour arises from mature cartilage.^{10,11} The extra skeletal chondrosarcomas are classified into well-differentiated, myxoid and mesenchymal forms, out of which myxoid and mesenchymal forms are more common.¹² Chondrosarcomas are graded into 3 histological groups based on mitotic rate, cellularity and nuclear size. Grade I chondrosarcoma is characterized by mild hypercellularity, large vesicular nuclei and small nucleoli with a five-year survival rate is 90%. Grade II tumours have moderately increased cellularity with a 5-year prognosis of 81% survival. Grade III is rarer than I or II and carries a 5-year survival rate of 43%.^{5,13}

Radiologically, a plain skull radiography can show only bone destruction and calcification. Computed tomography imaging can show the tumour as a soft tissue mass, it's extend and bone destruction with areas of nodular or plaque-like calcification. A more specific imaging technique in the diagnosis of chondrosarcoma would be a magnetic resonance imaging. This tumour shows low signal intensities on a T1-weighted image, high signals on a T2-weighted sequence with differential enhancement on post-contrast gadolinium T1-weighted sequences.^{5,8,11,14,15} The differential diagnosis to be kept in mind includes chondroma, osteoblastoma, osteochondroma, meningioma, osteosarcoma and chondroid chordoma.^{6,8}

The primary modality of treatment is surgical excision of the tumour. Radical resection of the tumour often results in functional/cosmetic abnormalities most often requiring a complex reconstruction.¹⁵⁻¹⁸ Adjuvant radiotherapy or chemotherapy are also considered for less-well-differentiated tumors to prevent local recurrence. They are also given for residual disease, and palliation.⁵ Proton

radiation therapy studies have shown excellent local control and better survival rates with base of skull chondrosarcomas after surgical debulking.^{7,8}

Local recurrence and distant metastasis are common, especially in 20% cases, commonest site being lungs. The most common cause of death in chondrosarcoma is uncontrolled local disease.^{6,14} Therefore, lifelong follow up is a must in case of head and neck chondrosarcomas.

CONCLUSION

Head and neck chondrosarcomas are rare, out of which nasal septal chondrosarcomas are even rarer. These tumours are locally aggressive and can present with symptoms mainly due to mass effect. This tumour is commonly seen in middle aged males. Radiological evaluation should be done where CT scan can show the extend of mass lesion with local bone destruction and calcifications. MRI gives a clearer picture of the tumour. Surgical resection is the treatment of choice for chondrosarcoma irrespective of the site. The diagnosis is confirmed by histopathological evaluation which also gives an idea about the prognosis and survival of the patient. Through our study we have clearly discussed how to properly evaluate a case of chondrosarcoma of nasal-septum, the management modality and also the importance of follow-up.

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