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Case Report

Chondrosarcoma of Nasal Septum-A Rare Case with Unusual Presentation

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Abstract

Chondrosarcoma usually arises in the pelvis, ribs or long bones but rarely may occur in the nasal septum. The nasal septum chondrosarcoma may extend to skull base, paranasal sinuses or orbit which is usually observed during recurrence. Radiological examination and histopathology plays an important role for its diagnosis and to assess the grade and extension of the tumor. Moreover, the morphological differentiation of low grade chondrosarcoma from chondroma and other tumors like metastatic renal cell carcinoma, chordoma may pose diagnostic challenge on histopathology. It is therefore essential to vigilantly examine the histopathological sections for subtle features for definite diagnosis of chondrosarcoma. The present case is therefore being reported of rare tumor of low grade chondrosarcoma of nasal septum in young female extending to skull base at initial presentation. The case also highlights the importance of close histomorphological examination of low grade chondrosarcoma to avoid diagnostic pitfalls and the importance of repeat biopsy if clinico-radiological suspicion is strong and thus avoiding further complications.
Keywords: Chondrosarcoma; nasal septum; low grade.

INTRODUCTION

Chondrosarcoma, a malignant chondroid tumor, comprises about 10-20% of primary bone tumors. It usually arises in the pelvis, ribs or long bones but rarely may occur in the nasal septum [1]. Males are more commonly effected with the usual age of presentation being 40 to 50 years [2, 3]. The nasal septum chondrosarcoma may extend to skull base, paranasal sinuses or orbit which is usually observed during recurrence [4, 5]. The presenting symptoms depend on the size and extent of tumor including nasal obstruction, discharg, diplopia, pain and facial swelling. Radiological examination and histopathology plays an important role for its diagnosis and to assess the grade and extension of the tumor. Moreover, the morphological differentiation of low grade chondrosarcoma with chondroma and other tumors like metastatic renal cell carcinoma, chordoma may pose diagnostic challenge on histopathology. It is therefore essential to vigilantly examine the histopathological sections for subtle features for definite diagnosis of chondrosarcoma. The present case is therefore being reported of rare tumor of low grade chondrosarcoma of nasal septum in young female extending to skull base at initial presentation. The case also highlights the importance of close histomorphological examination of low grade chondrosarcoma to avoid diagnostic pitfalls and the importance of repeat biopsy if clinico-radiological suspicion is strong and thus avoiding further complications.

CASE REPORT

A 32 year old female presented with right nasal obstruction, stuffiness and frontal headache for last three months. Laryngeal endoscopy revealed firm nasal mass in right nasal cavity involving posterio-lateral nasal wall and obstructing nasopharyngeal view (Figure 1A). Contast enhancing computerised tomography (CECT) neck showed heterogenously enhancing mass lesion of size 6x5x5 cm with multiple specs of calcification in right nasal cavity eroding nasal septum, ethmoid and sphenoid sinuses with extension towards skull base (Figure 1B). Bone scan was performed which showed increased tracer uptake in

skull. The patient underwent right nasal mass biopsy and fragmented tissue was obtained and histopathological diagnosis suggestive of inflammatory polyp was given. However in view of strong clinico-radiological suspicion of malignant tumor a repeat biopsy was done and final histopathological diagnosis of low grade chondrosarcoma (Grade I) was rendered. The sections showed tissue lined by pseudostratified ciliated columnar epithelium with sub epithelial tissue showing lobulated chondroid tumor. The tumor was having 1-2 cells per lacunae with nuclei showing mild atypia and coarse chromatin and at places it was permeating the bony bits. Mitotic figures were sparse (less than 1 per high power field) and necrosis was focal. (Figure 2 A, B) Immunohistochemistry of the tissue confirmed the diagnosis with vimentin and S 100 positivity of the tumor cells and Ki 67 index of 8-10% (Figure 2 C, D). The patient underwent resection of right nasal cavity, septum, nasal turbinates, sinuses and medial wall of right maxillary sinus with tumor via lateral rhinotomy. The patient was discussed in the institutional tumor board and was planned for external beam radiation therapy (EBRT). She received 54Gy/30 cycles to right nasal cavity by IMRT technique on 6 MV linear accelerator and she responded well to the treatment. On follow up repeat CT nose and paranasal sinuses reported no residual or recurrent lesion. The patient is on continuous follow up with no significant abnormality since last two years.

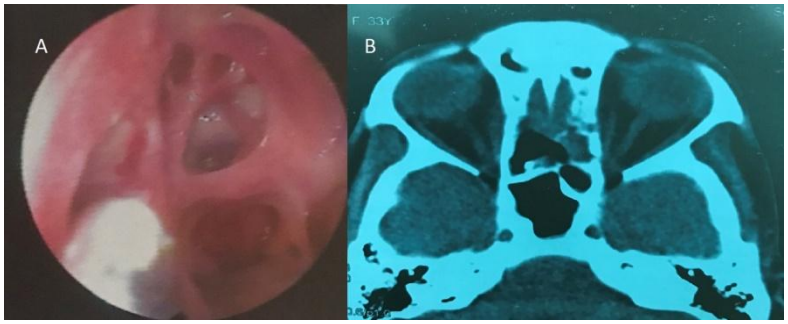

Fig-1: A) Laryngeal endoscopy showing firm nasal mass in right nasal cavity involving posterio-lateral nasal wall and obstructing nasopharyngeal view. B) CECT neck showing heterogenously enhancing mass in right nasal cavity eroding nasal septum, ethmoid and sphenoid sinuses with extension towards skull base

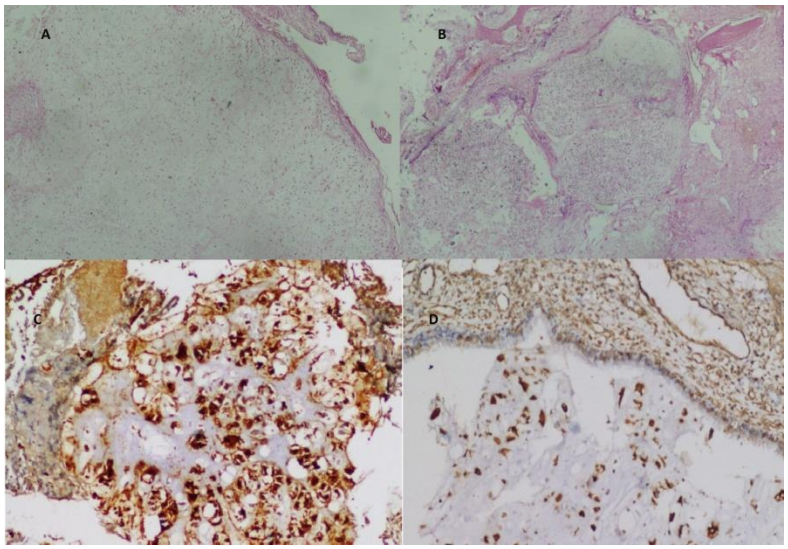

Fig-2: A) Biopsy of nasal cavity showing tissue lined by pseudostratified ciliated columnar epithelium with sub epithelial tissue showing lobulated chondroid tumor. (Hematoxyline eosin, x4). B) The tumor was having 1-2 cells per lacunae with nuclei showing mild atypia and at places it was permeating the bone (Hematoxyline eosin, x10). C, D) Immunohistochemistry of the tissue confirmed the diagnosis with vimentin and S100 positivity of the tumor cells

DISCUSSION

Chondrosarcoma is an uncommon tumor which rarely occurs in the nasal septum, sinuses, nasopharynx maxilla and mandible [6]. At times it may be difficult to suspect it in nasal septum due to generalised symptoms like nasal obstruction, facial pain or swelling and radiological examination may be necessary to know its extension to neighbouring tissues. It usually occurs in elderly males but the present case was reported in young female arising from nasal septum and involving the paranasal sinuses and extending to skull base at initial presentation. CT and magnetic

resonance imaging (MRI) are although informative but may be variable and should be carefully examined to know the intraorbital and intracranial extent of tumor with surgical margins [4, 7]. In our case the initial biopsy was diagnosed as inflammatory polyp but as radiological suspicion was strong for malignancy repeat biopsy was performed and diagnosis of chondrosarcoma was confimed. Thus the case highlights the importance of repeat biopsy if clinico-radiological suspicion is strong for malignancy as initial biopsy may sometimes be small or fragmented to misinterpret the low grade chondrosarcoma as unremarkable cartilage of nasal septum.

On histopathology, chondrosarcoma is lobulated cartilaginous tumor with increased cells per lacunae, anisocytosis and hyperchromasia and are graded on these features along with necrosis and mitosis. The important differential diagnosis which should be excluded are chondroma, adenocarcinoma or chordoma. However the close examination of the sections without physalipharous cells excludes chordoma, well circumscribed lobulated tumor with uniform cells and usually presence of single cell per lacunae without any permeation of tumor in surrounding tissue favours chondroma. Adenocarcinoma especially metastatic renal cell carcinoma lacks chondromyxoid matrix seen in chondrosarcoma. Cases have been reported in literature where nasal chondrosarcoma showed rare presentation of association with Maffucci syndrome or myxoid differentiation [4, 8].

The treatment of nasal chondrosarcoma requires surgical excision of the tumor which may be done either endoscopically or by an external approach including transnasal or lateral rhinotomy. The endoscopic resection is preferred if the tumor is of limited size without erosion of skull base and intracranial extension [9, 10]. In the present case the tumor was extensive and therefore resected by lateral rhinotomy approach. The chondrosarcoma is radioresistant tumor but may be subjected to radiotherapy if the tumor is incompletely resected due to extensive involvement as observed in the present case [10]. The prognosis of low grade chondrosarcoma is considered good if adequately treated but recurrence may occur after many years of primary diagnosis and therefore continuous follow up is necessary. The present case is also on continuous follow up and is disease free for last two years.

Conclusion

Nasal septum chondrosarcoma is a rare tumor and may be unusually present in females of younger age group with extensive involvement of paranasal sinuses and skull base. Radiological and histopathological examination is essential for its diagnosis and to know extension of the tumor. Vigilant histomorphological examination of low grade chondrosarcoma avoids diagnostic pitfalls and excludes chondroma and other similar tumors. Repeat biopsy may be important if clinico-radiological suspicion is strong for definite diagnosis and thus avoiding further complications.

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