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Sinonasal chondromyxoid fibroma: A rare tumour

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Abstract

Chondromyxoid fibroma (CMF) is a relatively rare benign bone neoplasm. CMF of the sinonasal tract is very rare. A 30-year-old male presented with nasal obstruction, right nasal mass with headache since two months. A Computed Tomographyscan showed locally aggressive hypodense, mildly enhancing lesion of size 10.3 cm x 5 cm x 5.1 cm involving right maxillary, ethmoid and frontal sinus with extension into premaxillary area and infratemporal fossa of the same side and into the nasopahrynx. Histopathology of biopsy revealed inflammatory polyp. The patient underwent a right medial maxillectomy with ethmoidectomy with excision of whole of the mass by lateral rhinotomy approach. A histological examination showed stellate cells in a chondromyxoid background with mitotic figures. It was provisionally diagnosed as Chondroblastic osteosarcoma. Immunohistochemistry confirmed the diagnosis of Chondromyxoid fibroma.

Keywords: Benign, Bone neoplasms, Chondromyxoid Fibroma, Craniofacial.

Introduction

Chondromyxoid fibroma is extremely rare tumour. It accounts for less than 1% of all primary bone neoplasms.(1) This distinct tumour is of cartilaginous origin and was first described by Jaffe and Lichtenstein in 1948.(2) Chondromyxoid fibroma is characterised by presence of variable amount of chondroid, fibromatoid and myxoid components. It is most frequently found in young adults of second and third decade of life in long bones of lower extremities and particularly arising from metaphysis.(3) It's occurrence in craniofacial bones is exceedingly rare. It is a slow growing tumour and develops symptoms over a period of months to years. Symptoms depend on the site and size of tumour. It is often mistaken for three other myxoid tumours: chondroma, chondroid chondroma, chondrosarcoma and have greater frequency of occurrence in the craniofacial skeleton.(4-5)

Here, we report a very rare case of sinonasal chondromyxoid fibroma with a diagnostic dilemma treated by surgical excision.

Case History

A 30-year-old male patient, farmer by occupation visited to ENT outpatient Department of Tertiary Health Care Hospital with a complaint of right sided nasal obstruction with mass protruding from right nasal cavity for 2 months (Fig. 1).

Fig.1: Right nasal mass protruding out through vestibule

He also noticed ipsilateral swelling over cheek, watering from right eye, Headache and anosmia. ENT examination revealed brownish crusted mass protruding out of right nostril with flared and tender ipsilateral ala and cellulitis over malar region. There was gross deviation of nasal septum to opposite side. On posterior rhinoscopy, there was a mass occupying whole of the nasopharynx covered with yellow slough. His ophthalmic check-up revealed normal vision and fundus examination in both eyes.

Histopathology report of biopsy which was done at some other centre was suggestive of inflammatory polyp. A Computed Tomographic (CT) imaging with IV contrast was done which showed locally aggressive hypodense, mildly enhancing lesion of size 10.3 cm x 5 cm x 5.1 cm involving right maxillary and ethmoid sinuses (Fig. 2) and also extending to frontal sinus, premaxillary area and infratemporal fossa of same side and nasopharynx (Fig. 3).

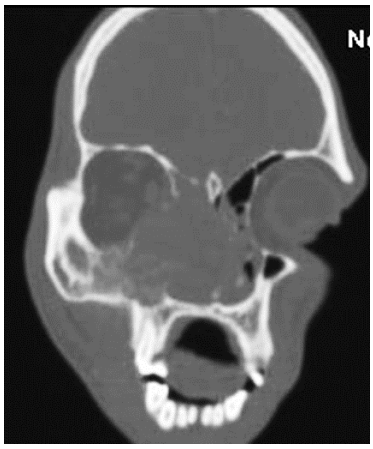

Fig. 2: CT PNS showing hypodense lesion occupying Right maxillary and anterior ethmoid sinuses

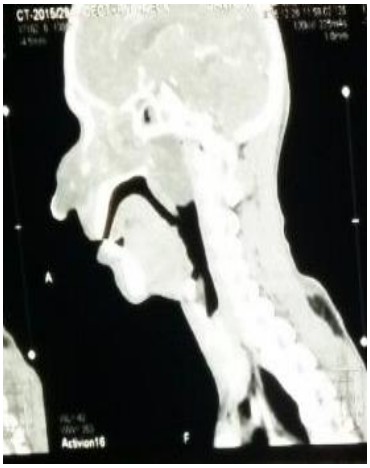

Fig. 3: Sagittal Section of CT PNS showing hypodence lesion extending to nasopharymx

There was subtle erosion of ipsilateral lamina papyracea. We did repeat biopsy of the mass nasal cavity and Histopathology report was consistent with the previous one. Histopathology report was not matching the extensive lesion clinically and radiologically with short history. So, we proceeded with Caldwel Luc's operation to get deeper biopsy, as the endoscope can't be negotiated through the huge mass to the nasal cavity. To our surprise, histopathology report was again inflammatory polyp.

Under general anesthesia, tumour was approached through Right Lateral Rhinotomy approach. Right medial maxillectomy with ethomidectomy was done to remove the tumour completely (Fig. 4).

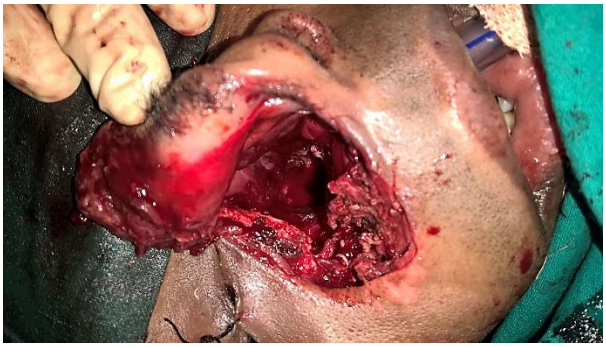

Fig. 4: Surgical excision of mass by Lateral Rhinotomy

Tumour extending at ipsilateral premaxillary and infratemporal region was also removed. Underlying bone was curetted well and surgical site was closed. Histopathologically, spindle cells in a chondrmyxoid storma arranged in lobules along with the osteoids with few areas of calcification was seen. The nuclei of Spindle cells were showing marked pleomorphism, hyperchromatism and mitosis. The chondrocytes were also showing nuclear pleomorphism. Provisional impression of pathologist was Chodroblastic tumour, most likely Chondroblastic osteosarcoma. To arrive to the final diagnosis, specimen was sent for Immunohistochemical analysis. It was positive for Vimentin, S-100, Smooth Muscle Actin (SMA) and was negative for Desmin, CD 31 and CD34.So, we confirmed the diagnosis by Immunohistochemistry as Benign Chondromyxoid Mesenchymal Tumour with no evidence of unequivocal invasive malignancy. In one year's follow- up, patient showed no clinical or radiological evidence of residual or recurrent disease.

Discussion

Chondromyxoid fibroma (CMF) is a rare benign tumour that accounts for 1% of all bone tumours.[1] Approximately 2/3rd of all cases of CMF occur in the metaphysis of long bones commonly affecting tibia and fibula. Craniofacial involvement is extremely rare. Mandible and maxilla are more commonly affected in this region. The World Health Organization defines CMF as "a benign tumour characterized by lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material".[6]

This tumour type has been described in neonates and adults, with a peak in second and third decade of life. There is a male predilection; but in cases of cranium and face bones involvement, women prevail (2:1).[7] Most common presenting symptoms depend on the site of involvement. Sinonasal CMF present with nasal obstruction, clival/ sellar lesions present with headache, sphenoidal/ parasellar lesions present with diplopia. Orbital/ zygomatic CMF presents most commonly with exophthalmos and lastly temporal/ occipital CMF is associated with deafness and otalgia during presentation.

There is no established radiological pattern for CMF because of its rarity but they can offer insight into diagnosis before intervention. CT scan shows radiolucent, lobulated, circumscribed lesion with sclerotic rim and cortical expansion or erosion. MRI findings are not clearly established because of low occurrence rate. It has low signals on T1 weighted and high signals at T2 weighted images similar to other cartilagenous tumours.

CMF is difficult to differentiate histologically as well as clinically. Histopathologically, CMF resembles to chondroblastoma and chondrosarcoma.[8-9] Misdiagnosis rate reported in literature is 18%.[8] It is very important to distinguish it from chondrosarcoma because management of these two entities differs. CMF is usually well-demarcated nodular lesion with myxoid lobules separated by thin, fibrous septa. Importantly, aCMF has a pauci-cellular centre rather than the uniform cellular arrangement observed in chondrosarcoma, and mitotic figures are exceptional.[6] In addition, the differential diagnosis includes fibrous dysplasia and chordoma. If the lesion shows significant atypia and mitotic figures diagnosis should be reconsidered. On immunohistochemistry, CMF is most commonly found to be positive for Vimentin, Smooth Muscle Actin, Desmin and S-100 (variably) and CD34.Generally, it is negative for pancytokeratin, carcinoembryonic antigen (CEA), and GFAP[10]. Sox9 is a transcription factor that was recently described as a master regulator of chondrogenesis. It plays a role in the early phases of chondrocyte differentiation.[11]

Complete surgical excision is the first line of treatment.[1] Because of the functional and cosmetic deformities that result from complete surgical resection a CMF of the craniofacial bones are best managed by curettage. Curettage is usually successful but increases the risk of recurrence[1] which is approximately 25%.[12] Such recurrence is usually local and malignant transformation is unlikely.[4] Role of radiotherapy in the management of CMF is controversial. Some authors recommend using radiotherapy for local relapses following surgical excision, particularly at the base of the skull.[13]

Conclusion

A sinonasal CMF is very rare and frequently misdiagnosed. Although CMF is a benign bone tumour, it should be differentiated from more aggressive malignant tumour. Patients who receive curettage have high recurrence rate. Periodic careful surveillance is necessary in such patients.

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