Chondromyxoid fibroma of nasal septum: A case report

Girija C and Bhavya J Menon

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Abstract
Chondromyxoid fibroma is an unusual tumour occurring in the metaphysis of long bones. It is a locally painful benign cartilaginous lesion with a high recurrence rate, not known to undergo malignant transformation. Involvement of cranial bones and Sino nasal region by this tumour is extremely rare. We present a case of chondromyxoid fibroma in a young male, arising from the nasal septum which is a very unusual site for this tumour.

Keywords: Chondromyxoid fibroma, metaphysis, aneurysmal bone cyst, microscopically

Introduction
Chondromyxoid fibroma (CMF), a neoplasm of cartilaginous origin along with myxoid and fibrous tissue, was first described by Jaffe and Lichtenstein in 1948 [1]. It accounts for less than 0.5% of all bone tumours with a predilection for lower extremities [2-4] and a slight male predominance [5]. These tumours are more common in the metaphysis of long bones. About 1-5% of Chondromyxoid fibroma cases are also reported to occur in the head and neck region [5, 6]. Common radiographic findings are an eccentric radiolucent defect without calcification, the adjacent cortex may be expanded thinned or even absent. Sometimes a sclerotic and scalloped rim may be seen. It is typically located in the metaphyseal region of long bones, and in some cases, it may invade the epiphyseal plate. Radiographic differential diagnosis of CMF includes non-ossifying fibroma and Aneurysmal Bone Cyst [7].

Case Report
A previously healthy, 24-year-old male presented with complaints of right-sided partial nasal obstruction of 10 months duration. He noticed a swelling in the right nasal cavity which was progressively increasing in size. There was no associated pain or bleeding from the nose. No other relevant past or family history. The patient was admitted to the Otorhinolaryngology department for surgery. On examination, there was a nasal mass arising from the upper part of the right side of the nasal septum and the roof of the nasal vestibule of size about 1.5x1cm which was firm in consistency. The surface appeared blackish brown in colour. There was no tenderness and it did not bleed on touch. Other general and systemic examination findings were within normal limits and had nil significance. No focal neurological deficits were identified. A CT scan of the Paranasal sinuses showed a well-defined, soft tissue density measuring around 1.3x0.8 cm in the right anterior nasal cavity causing partial obstruction, abutting the nasal septum and vestibule. At Diagnostic Nasal Endoscopy (DNE), the nasal mass was seen arising from the roof of the right nasal vestibule with a wide base, surface skin covered and appeared pigmented.

Surgical excision of the mass was done under GA in March 2021 and the specimen was sent for histopathological examination.

Grossly it was a polypoidal skin-covered nodular soft tissue mass of size 2x1.5x0.8cm. The Cut section was grey-white, firm with areas of focal cystic change and multiple tiny specs of calcifications. Microscopically the tumour was well-circumscribed, lobulated, covered by normal stratified squamous epithelium, bearing benign mature cartilage. The interlobular areas showed fibrovascular bundles and multinucleate osteoclast-like giant cells. The lesion was moderately cellular at the periphery composed of stellate to oval cells scattered in a chondromyxoid matrix along with a few multinucleate osteoclast-like giant cells.
Towards the centre, it was hypocellular to a cellular. No mitoses or necrosis was seen within the tumour. No cellular atypia or infiltrative margins were identified. All margins of resection were free of tumour.

**Discussion**

Till date, there have been 21 reported cases of Sino nasal CMF presenting in the nasal bones and paranasal sinuses [8, 9]. The symptoms of CMF vary depending on the location and the size of the tumour. It includes diplopia, facial pain, exophthalmos, neuralgia, dysarthria, epistaxis, nasal congestion, headache, bony swelling, or persistent chronic rhinosinusitis etc, not relieved by medical management [10]. These symptoms may be generally of slow onset and can vary according to the exact location of the tumour and the structures that are compressed or invaded. Because it occurs so rarely, CMF of the head and neck is often misdiagnosed. Imaging studies can be helpful in making the diagnosis and in planning management. High-resolution CT is preferred for defining the relationship between the tumour and surrounding bone, and magnetic resonance imaging can be very helpful in identifying the extent of Dural or intracranial involvement if any [11]. In our case, the tumour was composed of fibrous tissue and stellate cells studded in a chondromyxoid matrix, a few differentials were thought of and ruled out. A possibility of a cemento-ossifying fibroma was considered because of the spindle stromal cells, but the absence of basophilic, mineralised cementum and Sharpey’s fibres ruled out its possibility. Low-grade chondrosarcoma was ruled out due to the lack of diffuse hypercellular areas, cellular atypia and radiological evidence of popcorn or punctate calcifications and also considering the young age of the patient. Chondroblastoma was also ruled out due to the absence of chicken wire calcification in the lesion. A possibility of chordoma was ruled out due to the absence of physaliferous cells. Considering the age of the patient, chondromesenchymal hamartoma was also ruled out as they occur in patients in less than 3 years and it usually shows hyaline cartilage. Another rare and congenital bone tumour that can occur in nasal bone considered was osteochondromyxoma [12]. But the possibility was ruled out as it is usually associated with Carney’s complex and it is known to erode the bone and frequently extend to the soft tissue. Our lesion was well confined and had smooth polypoidal architecture.

Since the histomorphology findings of the lesion were not compatible with any of the above differentials, a diagnosis of the Chondromyxoid fibroma of the nasal septum was considered. What makes this case special is the rarity of chondromyxoid fibroma arising in a nasal septum.

Surgery is the most widely accepted modality of treatment for CMF the options being curettage and a wide en bloc excision. Curettage is used less often because of the relatively high risk of recurrence (12.5 to 25%) and the higher risk of cerebrospinal fluid leak or meningitis. Complete excision with wide margins is associated with a lower recurrence rate, but also with considerably greater morbidity. In the head and neck, complete resection is often difficult in such cases, a partial resection with long-term surveillance is an acceptable plan [13].

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**Fig 1A:** Low power view of the lesion from the nasal septum. Overlying normal cartilage noted with nodular lesion underneath.

**Fig 1B:** Overlying stratified squamous epithelium with underlying lesion with lobulated architecture.

**Fig 2:** Lobules with hypocellular centres and hyper cellular periphery.

**Fig 3:** Variable myxoid to chondromyxoid stroma, having stellate cells and spindle cells.
Fig 4: Stellate cell-rich areas in chondromyxoid fibroma

Fig 5: Variably myxoid to chondroid stroma with various stages of cartilaginous development

Fig 6A: Areas showing spindle cells and multinucleate giant cells.

Fig 6B: Bipolar and multipolar spindle cells with multinucleate giant cells at the periphery of the CMF

Conflict of Interest
Not available

Financial Support
Not available

References

12. Pathology outlines.


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