Chondrosarcoma of the maxilla. Case report

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Abstract
Chondrosarcoma is a rare malignant neoplasm of the jaws. The diagnosis of this lesion is mainly based on the histopathological appearance. A case of chondrosarcoma of the maxilla is reported. The origin of this rare tumour in the maxilla, a bone with exclusive membranous ossification, is discussed.

Key words: Chondrosarcoma, maxilla.
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Introduction
Chondrosarcomas are malignant tumours arising from cartilage cells that tend to maintain their essentially cartilagenous nature throughout their evolution. These neoplasms mostly occur in the long bones, the pelvis, and the ribs. They are rare in the head and neck region which accounts for less than 1 to 2 per cent. Most chondrosarcomas of the head and neck region occur in the maxilla; others are found in descending order of frequency in the body of the mandible, the ramus, the nasal septum, and the paranasal sinuses.

The aetiology of these tumours is unknown. However, they are formed from cartilage in tissues not normally harbouring cartilage or, secondly, from the cartilage cap of exostosis or enchondromas. Vestigial rests of multipotential differentiation of mesenchymal cells may be the forerunner.

Chondrosarcomas of the head and neck have been reported in patients ranging in age from 17 months to 75 years. The peak age of incidence is the third to the sixth decade. There does not seem to be a sex predilection. The tumours show a less aggressive course when found in the long bones rather than when found in the head and neck as the latter have a greater rate of growth, recurrence, and metastasis. They usually present as painless swellings.

A case of chondrosarcoma of the premaxilla is presented. The clinical and histopathologic features are discussed.

Case report
A 24 year old female reported to the Outpatient Department of the Government Dental College and Hospital, Trivandrum, India, with complaints of a painless swelling in the palatal region. The patient had noticed the swelling six months earlier and it had gradually increased to its present size. There was difficulty in mastication.

On examination, a firm, non-tender swelling of 40 x 60 mm in dimension was noted in the premaxillary region extending bilaterally from the maxillary incisor region to the premolar region (Fig. 1). A radiograph of the maxilla showed a radiolucent lesion with well circumscribed borders (Fig. 2). Histopathological examination revealed cartilage cells showing considerable variation in size with binucleated cells being common. These nuclei were often larger than normal and some mitotic figures were noted. The tumour matrix showed a variation in staining reaction which is consistent with chondrosarcoma (Fig. 3).

The patient was referred to the Regional Cancer Centre for further management. Total maxillectomy was advised, but the patient did not report back for further treatment or follow-up.

Discussion
A review of the literature reveals that chondrosarcoma of the maxillofacial region is extremely rare. The neoplasm has an aggressive local behaviour as well as a highly metastatic potential, and the long-term prognosis is extremely poor. Chondrosarcoma
of the head and neck occurs most often in the third to the sixth decades of life. Its histogenesis is still unclear. However, observations suggest that the tumour may originate from cell populations in the course of the early differentiation stage of chondrogenic linkage as compared with conventional chondrosarcoma. Clinically, there are no signs and symptoms other than swellings or masses. Radiographically these lesions appear as osteolytic, radiolucent shadows with ill-defined, ragged borders. Diagnosis can only be established by histopathological examination. Microscopically, the lesions show a proliferation of hyaline cartilage formed by a sarcomatous stroma containing stellate,
spindle-shaped and round cells. The cells show a greater degree of pleomorphism and atypia. Studies have revealed that chondrogenic tumours of the jaws are often more malignant than benign. Benign tumours that have shown a sudden increase in growth have undergone malignant transformation.

Chondrosarcomas are radio-resistant lesions. Radical surgical excision is the method of choice as this has demonstrated the best chance of effecting a cure. Radiation and chemotherapy has been used as a supplement to surgery but have been ineffective. Sato et al. believe that recurrence of chondrosarcoma is frequent because of the complicated location which does not allow complete surgical excision of the lesion. The prognosis for patients with chondrosarcoma appears to be related to the location of the tumour, the adequacy of the primary surgical resection, and the histological grade of the neoplasm.

References

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