



Chondrosarcoma of the Maxilla-Recurrent

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Abstract

An uncommon case of recurrent chondrosarcoma of the maxilla is presented. A brief discussion on the radiologic and histologic presentation of the tumor and the treatment modalities of this unusual tumor is discussed.

Key words

Chondrosarcoma, Craniofacial, Maxilla

Introduction

Chondrosarcoma is an uncommon malignant neoplasm of cartilagenous origin devoid of tumor osteoid with the evidence of fully developed cartilagenous structures. It is an extremely rare primary tumor of head and neck with less than 10% of the cases occurring in craniofacial region. The chondrosarcoma of the craniofacial region may arise from any bone, cartilage or soft tissue structures. In head and neck, the most common sites of origin are the maxilla, mandible, nasal septum, sphenoid sinus and the ethmoid sinus (1). We, hereby, present a rare case of recurrent chondrosarcoma of the maxilla.

Case Report

A 45-year-old man was admitted with a bony swelling on the right side of the hard palate. He was operated for chondrosarcoma of nasal cavity, by bilateral lateral rhinotomy approach 6 years back and his histopathological report was documented as well differentiated chondrosarcoma grade-I with all margins negative for the tumor.

There was no history of pain, nasal discharge or obstruction, epistaxis or loosening of teeth, diplopia, diminished vision or headache. Physical examination revealed a diffuse bony swelling (1 cm × 1 cm) on right side of hard palate with intact palatal

mucosa. Anterior rhinoscopy revealed a smooth, pinkish mass on the lateral wall of the right nasal cavity, which was bony hard, non-friable in probe test. The bony septum was totally absent because of the previous surgery. Posterior rhinoscopy and rest of the ENT examination and cranial nerves examination was normal. Routine investigations were within normal limits. CT scan (Fig. I) of the paranasal sinuses revealed a bony mass arising from medial wall of the right maxillary antrum extending to the alveolar margin. Maxillary sinus mucosa was normal. There was no orbital, zygomatic, choanal or nasopharyngeal spread. Posterior part of the nasal septum was missing due to previous surgical intervention.

The patient was taken for surgery as a case of recurrent chondrosarcoma and the wide field radical surgery (inferior maxillectomy/infrastructural maxillectomy) was done under general anaesthesia. The post-operative period was uneventful. The histopathology of the resected specimen revealed well-differentiated chondrosarcoma with all resected margins free of the tumor (Fig. II).

Discussion

Unlike the expanding high grade chondrosarcoma of the long bones presenting with excruciating pain, the

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Fig 1: CT Scan of paranasal sinuses revealed a bony mass arising from medial wall of the right maxillary antrum extending to the alveolar margin

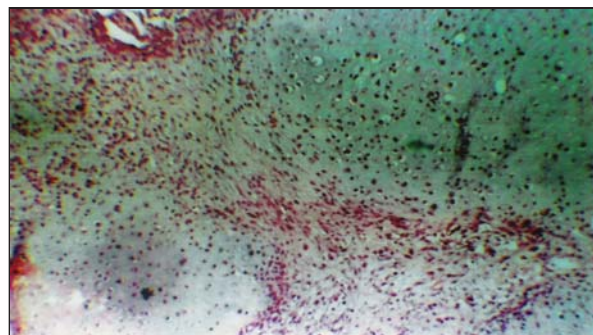


Fig II: Histopathology of Chondrosarcoma (H&E-10X)

chondrosarcomas of head and neck tend to be painless on presentation. Less than 10% of all cases of chondrosarcoma involve the craniofacial region, accounting for less than 2% of all head and neck tumors. It is usually found in adults between 3rd and 6th decade of life, although, the youngest patient reported is 16-month old baby and the oldest one a 74-year old man. The male to female ratio is 1.2:1 (1).

The most common clinical finding is painless swelling, expansion of the buccal and the lingual plates, premature eruption or exfoliation of teeth. The mass is usually rapidly growing and covered with mucosa which can ulcerate and there can be pain at later stages. Rarely there can be lymph node involvement. Headache, blurred vision and proptosis, diplopia and facial swelling are less frequent findings. It can also cause nasal stiffness, nasal discharge or epistaxis. The chief complaint of an edentulous patient may well be that of an ill-fitting denture (2, 3, 4). Myxoid chondrosarcoma, although rare in head and neck region, when present in children with swelling of face, reaffirm the importance of considering sarcomas or other neoplastic lesions in the differential diagnosis of progressive nasal obstruction in children (5).

The pathogenesis and biologic behaviour of the chondrogenic tumors is not fully understood, but it is evident that these lesions represent a spectrum from benign chondrosoma to the malignant chondrosarcoma, through all degrees of intermediate type (2). The exact origin of chondrogenic sarcoma in head and neck is controversial. It may be induced by irradiation, from pre-existing paget's disease of bone or in association with fibrous dysplasia and the solitary bone cyst or it may

arise from the vestigial cartilagenous rests. In pre-maxilla and maxilla, these cell rests are quite possible because of the proximity of chondrocranium throughout the fetal development. In mandible, the lesion may arise from the coronoid or condyloid process, from mental symphysis or from remanants of Meckel's cartilage (2, 3). Whereas, some believe that the chondrosarcoma can arise de-novo from osseus tissues without the presence of cartilagenous rests (4). Some authors believe that mandible is more common a site for chondrosarcoma than maxilla (3). While Terezhelmy (2) suggested that the tumor is found in equal frequency in both arches, Huvos reported maxillary predilection (6).

The radiological pattern of chondrosarcomas is variable (3). It includes single or multiple radiolucent areas. These lytic changes are prominent in more advanced cases. Other findings are opacification of air spaces, a densely calcified bone mass and root resorption (2). Also, it may reveal ground glass appearance or a sunburst appearance. Cortical destruction occurs late in the course of disease and periosteal bone formation is often limited (3). Some authors have reported a uniform widening of periodontal membrane space. In late stage disease, the primary lesion may penetrate the cortical plate and extend into adjacent soft tissues, resulting in a fuzzy soft tissue, peripheral shadow radiologically (4).

Histologically, chondrosarcoma continues to be defined as a malignant tumor composed of fully developed cartilage without tumor osteoid, being directly formed from a sarcomatous stroma. Myxoid changes, calcification and ossification may be seen. Evan's and co-workers have attempted to associate the histologic grade (Grade



I to III) of chondrosarcoma with the ultimate biologic behaviour of the tumor, depending upon the cellularity, nuclei size, presence of mitotic figures, multinucleation, spindle cell formation and mineralization in the form of osseous development at the edge of the cartilagenous lobules (7). In head and neck, the largest percentage of chondrosarcomas has been reported as grade-I (8).

Clinically, the chondrosarcomas of jaws are relatively invasive and destructive, with great tendency to recur unless adequately treated. Even benign lesions, following recurrence, are often found to be more cellular or to have already turned frankly malignant. Hence, all cartilagenous tumors of the jaws, benign or malignant, should be radically excised with a portion of the normal tissue to avoid recurrence (2, 3). The reported patient was showing the grade-I chondrosarcoma with margins free of tumor histologically when was operated first time, but even then, there was recurrence of the tumor.

The chondrosarcomas of the maxilla are classically treated by radical surgery with radiotherapy being used as an adjunct or a form of palliative treatment for recurrent lesions. These lesions are radio-resistant and therefore radiotherapy is not generally recommended as a primary modality (3, 9). Chemotherapy is sometimes used for palliation and so is the role of radioactive sulfur (S^{35}) (3). Electrocoagulation may also be an alternative to achieve cure (10).

The prognosis of the chondrosarcoma of the jaws is disappointingly poor as compared to that of long bones. The cause of death is usually by direct extension into the base of skull, and also through distant metastasis, chiefly to lungs and bones (7). The prognosis is good for low and intermediate grade chondrosarcomas. They are considered to be more dangerous with worse prognosis

than osteosarcomas of jaw. Specially the maxillary (0.7% of whole body tumor) and mandibular locations of the tumor has documented inferior prognosis (10).

The tumor involvement at the resected margins is the only other poor prognostic sign. The overall 5-year survival for low grade chondrosarcomas after complete resection is between 55-75% (1).

The reported patient has done well after 6 years of previous surgery and despite recurrence, could be managed by radical surgery and is doing well on follow-up for the last more than 6 months.

References

1. Lone Shafkat A, Mir S, Mohd L. Chondrosarcoma of the paranasal sinuses. *JK Science* 2003; 5 : 124-25.
2. Terezhelmy GT, Bottomley WK. Maxillary chondrogenic sarcoma : Management of a case. *Oral Surg* 1977; 44 : 539-46.
3. Greer RO, Rohrer MD, Young SK. Non-odontogenic tumors. In: Thewley SE, Panje WR (eds.). *Comprehensive Management of Head and Neck Tumors*. WB-Saunders Company, Oxford, 1987. pp. 1510-59.
4. Greer RO, Mierau GW, Favara BF. Tumour of head and neck in children. New York, Prager Publishers, 1983. pp. 125.
5. Jorg S, August C, Stoll W, Albery J. Myxoid chondrosarcoma of the maxilla in a pediatric patient. *Eur Arch Otorhinolaryngol* 2006; 263 : 195-98.
6. Huvos AG. Bone tumors : Diagnosis, treatment and prognosis. WB Saunders Co, 1979. pp. 5.
7. Evans HL, Ayala A, Romsdahl NM. Prognostic factors in chondrosarcoma of bone. A clinicopathologic analysis with emphasis on histologic grading. *Cancer* 1977; 40 : 818.
8. Chaudhary AP, Robinovitch MR, Mitchell DF, Vickers RA. Chondrogenic tumors of the jaw. *Am J Surg* 1961; 102 : 403.
9. Sonkhya N, Sharma N, Bhuie HS, Chhangani DL. Chondrosarcoma of the nasal septum. *Ind J Otolaryngol Head Neck Surg* 1991; 43: 150-51.
10. Biswas S, Mondal P, Saha S, Bera SP. Chondrosarcoma of the jaw. *Ind J Otolaryngol Head Neck Surg* 2005; (Special Issue No I): 294-98.

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