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Chondrosarcoma of the maxilla

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Abstract
A patient was referred to us with a mass in her upper jaw. This was diagnosed to be a chondrosarcoma of the maxilla. The mass was removed surgically. We present the case here and discuss the salient features.

Introduction

Chondrosarcoma of the maxilla is a rare entity. Nonetheless it is one of the most common primary bony tumour afflicting the maxilla (Batsakis, 1987). Of all chondrosarcomas about 10 per cent are seen in the maxillo-facial area (Berktold *et al.*, 1984).

In most published series only a small number of cases are reported. In view of the limited number presented many features of the disease are not clearly understood and contradictory views appear in the literature regarding its behaviour (Ajagbe *et al.*, 1985).

Maxillary chondrosarcoma most commonly present as a mass, the anterior alveolar area being involved in many cases. Pain is complained of by less than half of the patients initially. Other features which can be associated with the disease are nasal obstruction, epistaxis, paraesthesia accompanied by or without dental symptoms. Large tumours may invade the cranial nerves. Regional lymph node involvement is rare, though metastasis may occur late in the lungs (Gallagher and Strome, 1972). In most cases the tumour grows slowly and the disease remains locally invasive for a considerable period of time (Garrington and Collett, 1988).

We present a case here which illustrates many of the important features of the disease.

Case report

In October 1990, a 52-year-old lady was referred with a six-week history of a mass over the left upper alveolus. The mass caused displacement of her partial upper denture and considerable discomfort. However, the mass itself was painless. There was grade 3 mobility of the teeth in the left maxilla. Clinical

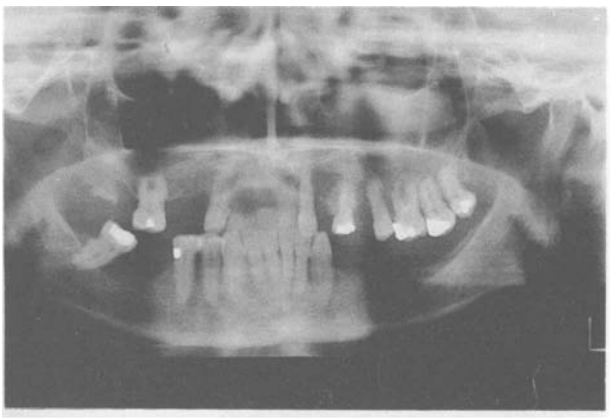

Fig. 1

Orthopantomogram: Left antral mass showing areas of bone destruction interspersed with areas of calcification, especially in the left canine and pre-molar region.

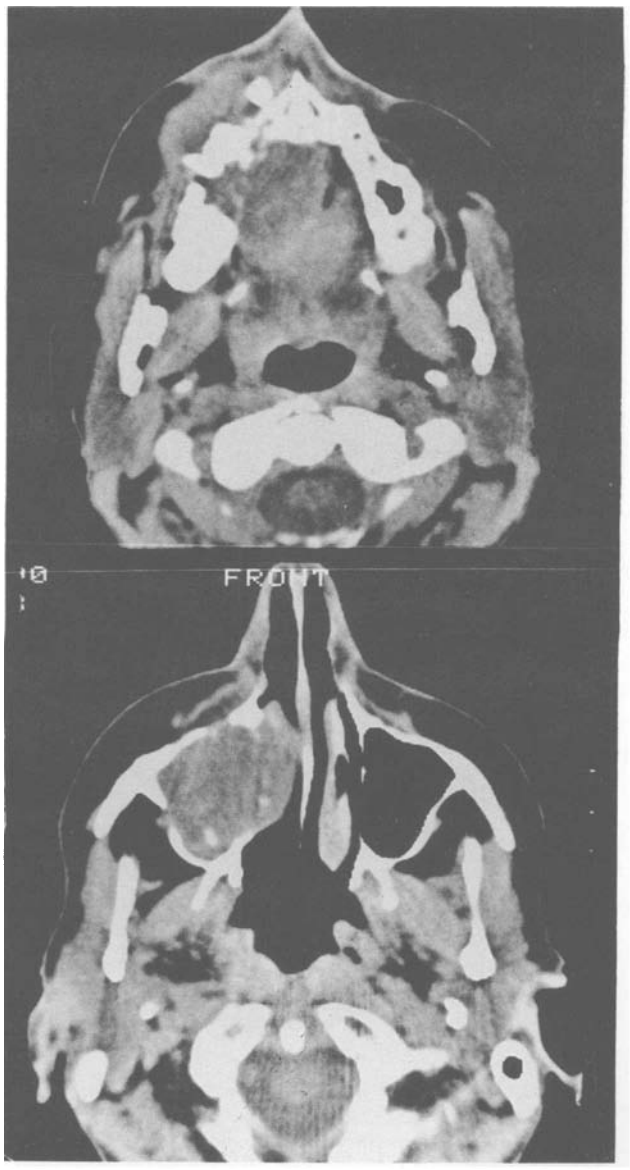

Fig. 2

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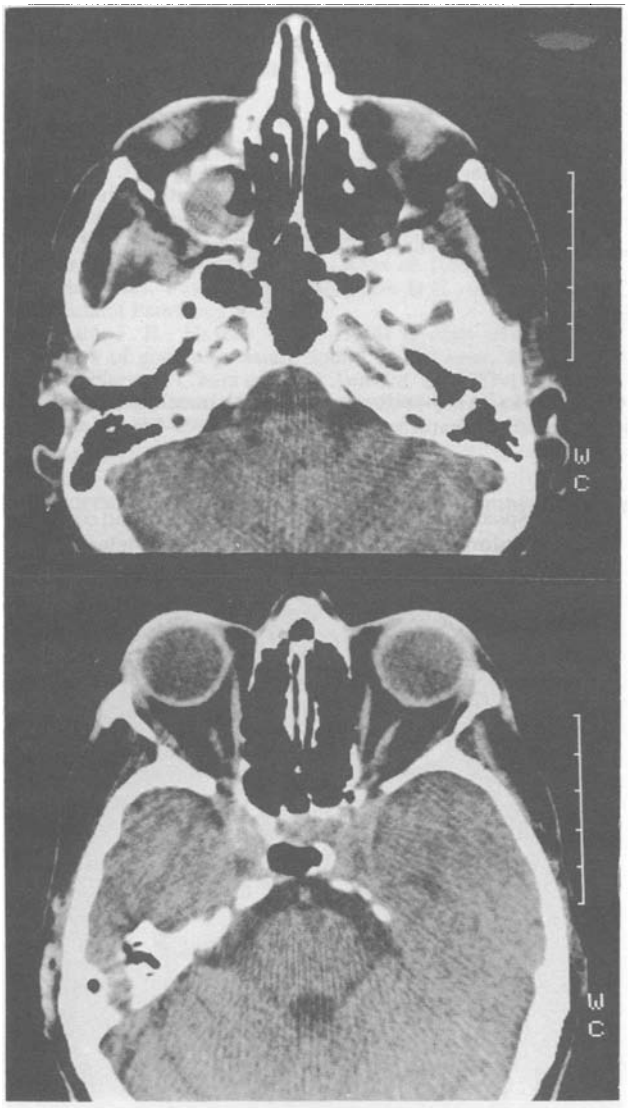

FIG. 3

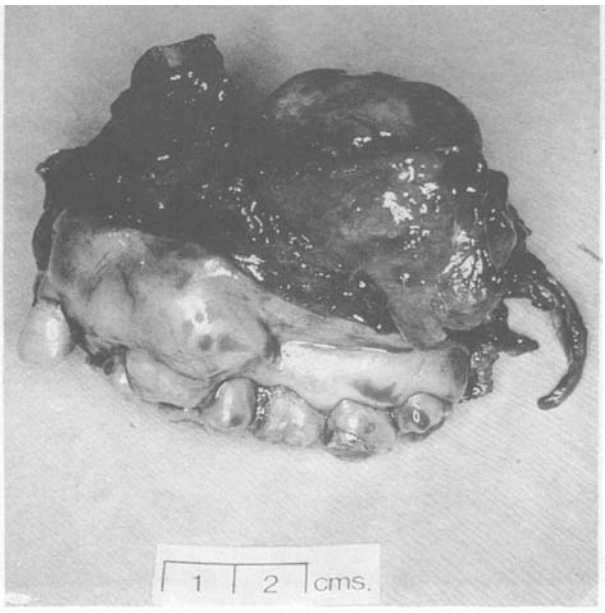

FIG. 4

Excised surgical specimen of the left hemi-maxilla showing a microscopically well demarcated tumour.

examination revealed a large swelling in the 1–6 region flattened by the denture and extending into the palate. There was no evidence of cranial nerve involvement and no clinical involvement of cervical lymph nodes. An orthopantomogram showed areas of osteolysis interspersed with areas of calcification (Fig. 1). The CT scan revealed an extensive soft tissue mass of the left alveolus and maxilla causing bone destruction anteriorly and medially and extending into the nasopharynx (FIg. 2). There was no significant posterior or lateral tumour extension and the orbit appeared clear of any tumour (Fig. 3).

An incisional biopsy was reported as a chondosarcoma of the maxilla and a left partial maxillectomy was performed without delay.

The maxilla was approached via a Weber-Ferguson's incision with blepharoplasty extension. The tumour was removed en-bloc after making suitable bony cuts through the hard palate, infra-orbital floor, frontal process of maxilla and the zygomatic buttress (Fig. 4). A temporary obturator was constructed to obturate the resulting surgical defect. Histological examination confirmed a complete local removal of the tumour.

Histopathology of the resected specimen revealed a well defined lobulated cartilagenous tumour within the maxilla, 3.5 cm in maximum dimension. The tumour had a shiny grey-white lobular cut surface with cystic areas associated with mucoid material and flecks of calcification.

Multiple blocks from the tumour showed a chondrosarcoma composed of cartilaginous cells with plump nuclei, increased cellularity, enlarged grotesque nuclei, multiple nuclei and occasional mitotic figures. The bulk of the tumour was fairly well differentiated (Grade 1) with mild to fairly obvious morphologic deviations (Fig. 5), but in a few areas anaplasia was marked with barely recognizable cartilage cells (Fig. 6). There were foci of calcification and ossification (Fig. 7). The tumour had eroded the cortex of the maxilla and ulcerated the buccal mucosa.

Her post-operative period was uneventful and she was well enough to be discharged home on the seventh post operative day. She is now being regularly followed-up in the Outpatient clinic.

Discussion

Various factors make it difficult for chondrosarcoma of the maxilla to be diagnosed with certainty. Conventional radiography may be of limited help in the early stages of the disease. This is due to the fact that the lesion may not show detectable radiological changes early on and even if it does, considerable overlapping and superimposition in this area may prevent detection (Cohen *et al.*, 1984).

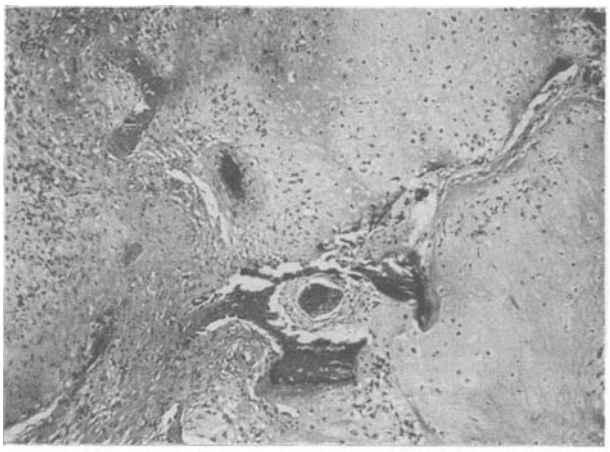

FIG. 5

This photomicrograph shows a well differentiated area of chondrosarcoma with lobular margins, mild cytologic atypia and focal enchondrial calcification.

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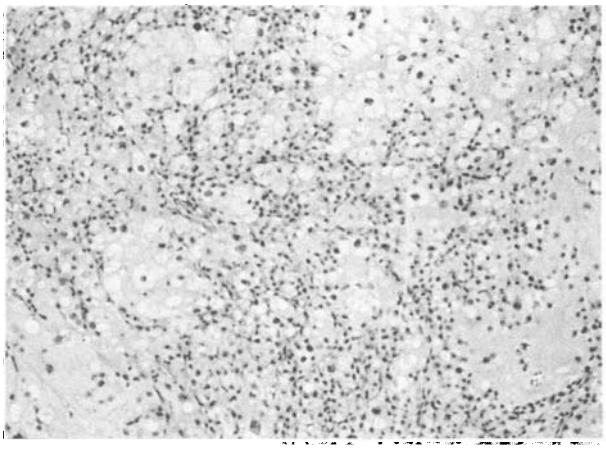

FIG. 6

Chondrosarcoma in this photomicrogram is from a grade 3 poorly differentiated area showing extreme cellularity and atypia of cartilage cells which have lost their normal grouping.

The common radiographic appearance is that of an osteolytic lesion, sometimes showing areas of speckled calcification. The presence of calcification indicates the probability of well differentiation of the tumour. The tumour may occasionally be densely calcified in which case X-ray may allow a 'sun-ray effect' (Sato *et al.*, 1977).

A CT scan with contrast enhancement can be invaluable in demonstrating the extent of the disease. Cohen *et al.* (1984) showed that the soft tissue component and extension into the surrounding structure which is poorly demonstrated by conventional radiography can be visualized by a CT scan with a high degree of accuracy.

Histological misdiagnosis of chondrosarcoma is not uncommon. Histologically, it is often difficlt to differentiate chondroma and chondrosarcoma, particularly cellular chondroma and low grade chondrosarcoma (Zachariades *et al.*, 1987). However, the occurrence of chondrosarcoma is twice as common as its benign counterpart in the craniofacial region and Batsakis (1987) emphasized that any cartilaginous lesion in this area should be evaluated cautiously.

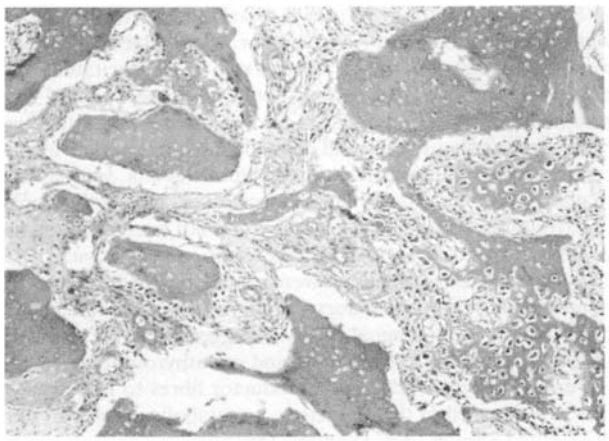

FIG. 7

Some areas in the tumour shows calcification and bone formation within malignant cartilage.

There is a general consensus that radical surgical excision with a wide margin of healthy tissue offers the only hope of eradicating the disease. Chondrosarcoma is usually very radioresistant. Radiotherapy and chemotherapy are considered ineffective therapeutically. Paddison and Haenes (1971) reported a good response to radiotherapy. However, in a review, Cohen *et al.* (1984) point out that most authors consider this disease to be radioresistant. Radiotherapy can be useful in the palliation of tumours considered to be beyond the scope of surgery.

Prognosis depends on histological grading, anatomical location and adequacy of surgical ressection.

Because the disease is notorious in being prone to local recurrence, long-term follow-up is vital. The benefit of a well obturated maxillary defect is that the whole surgical field is available for direct inspection.

Key words: Maxillary neoplasms, chondrosarcoma

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