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Chondrosarcoma of the parapharyngeal space

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Abstract
Chondrosarcoma is rarely found arising in the head and neck region. An unusual case arising in the parapharyngeal space in a male is reported and the differential diagnosis, pathology and treatment are discussed.

Introduction

Chondrosarcoma is a relatively rare malignant tumour most frequently affecting the pelvis, sternum, scapula, and long bones. Head and neck chondrosarcoma represents a small percentage of the overall incidence of this tumour. Arlen *et al.* (1970) described 18 cases involving the head and neck, but none of them was arising in the parapharyngeal space. McIlrath *et al.* (1963) reviewed the cases of 101 patients with parapharyngeal tumours seen at the Mayo Clinic from 1938 through 1958, but no chondrosarcoma was found.

This case report describes an extraskeletal chondrosarcoma arising in the parapharyngeal space in a 36-year-old patient.

Case report

A 36-year-old male was admitted for investigation of pain and swelling beneath the jaw on the left side which had been present for five months. On physical examination a firm swelling was noted beneath the angle of the left mandible and a bulging of the lateral pharyngeal wall into the oropharynx. The cranial nerves were normal.

A CT scan showed a solid lesion 5 cm diameter involving the medial pterygoid muscle anteriorly and laterally having unclear margin with parotid gland. There was no bone destruction and no prominent calcification within the lesion. Irregular enhancement was noted following the administration of intravenous contrast (Fig. 1). An isotope scan showed excess uptake of Ga^{67} in the tumour. Chest X-ray, routine haematological and biochemical tests were normal.

The external surgical approach was favoured for excision of the tumour. The facial nerve was exposed, and the vessels of the carotid sheath were identified. Access to the parapharyngeal space was improved by the surgical removal of the submandibular gland and a portion of the tail of the parotid gland. Grossly the material removed had a cartilaginous consistency (Fig. 2). The histological diagnosis of chondrosarcoma was made and post-operative radiotherapy of 6,000 cGy over six weeks was given. The patient has been followed up for six months without evidence of recurrence.

Pathology

Gross examination showed a tumour measuring $5 \times 3 \times 2$ cm, composed of lobulated, firm, grey-white tissue. On microscopic examination, the tumour was made up of vaguely defined nodules most of which had a matrix of hyaline cartilage. While few had fully differentiated cartilage cells in capsules, the greater majority had embryonal chondroblasts in the form of small rounded cells with one or two nuclei (Fig. 3). The greatest cellularity was located toward the periphery of tumour lobules. In these foci, the nuclear size was greater and the tumour cells showed numerous mitoses (Fig. 4). These lesions fulfilled the criteria for the diagnosis of chondrosarcoma Grade III as suggested by Evans *et al.* (1977).

Discussion

The clinical features of this case were those of a parapharyngeal tumour confirmed by CT scan. Tumours of the parapharyngeal space are similar in their manifestations, but they are remarkably different in their pathological features (McIlrath *et al.*, 1963). The symptoms were not specific for diagnosis. CT plays an important role in delineating the extent of the tumour with visualization of bony destruction, associated soft-tissue mass, and calcification, information which is extremely important for surgical and therapeutic planning (Hertzanu *et al.*, 1985). There is a relationship between the density of calcification in chondrosarcoma and the degree of malignancy. High-grade tumours are frequently associated with large areas of non-calcified tumour (Rosenthal *et al.*, 1984). Rosenthal *et al.* (1984) observed a strong correlation between the radiographic grade of osseous margins and the histological grade of the tumour. Infiltrative margins are considered to be characteristic of high-grade malignancy. In our case, the tumour was associated with little calcification and showed undefined boundaries between tumour and soft-tissue, but bone destruction was not seen. The diagnosis was made by histological study with microscopic features being typical for chondrosarcoma.

Chondrosarcoma is a malignant neoplasm of uncertain histogenesis which most commonly arises in the long bones and ribs. Ten per cent arise in the maxillo-facial area (Myers and Thawley, 1979), but most of them arise from the cartilaginous portion of the nasal skeleton. To our knowledge, only two cases (Wirth *et al.*, 1943; Watanbe *et al.*, 1988) of extraskeletal chondrosarcoma arising in the parapharyngeal space have previously been reported. Fu and Perzin (1974) have suggested that benign and malignant cartilaginous tumours probably arise from remnants of the embryonal cartilaginous skeleton which escape resorption during endochondral ossification. Extraosseous chondrosarcomas have the same histological features as chondrosarcomas of bone, and usually arise *de novo* rather than from pre-existing benign chondromas (Stout and Verner, 1953).

According to Fu and Perzin (1974) the prognosis of chondrosarcomas depends upon: (1) the location and extent of the lesion; (2) the adequacy of the surgical therapy; and (3) the

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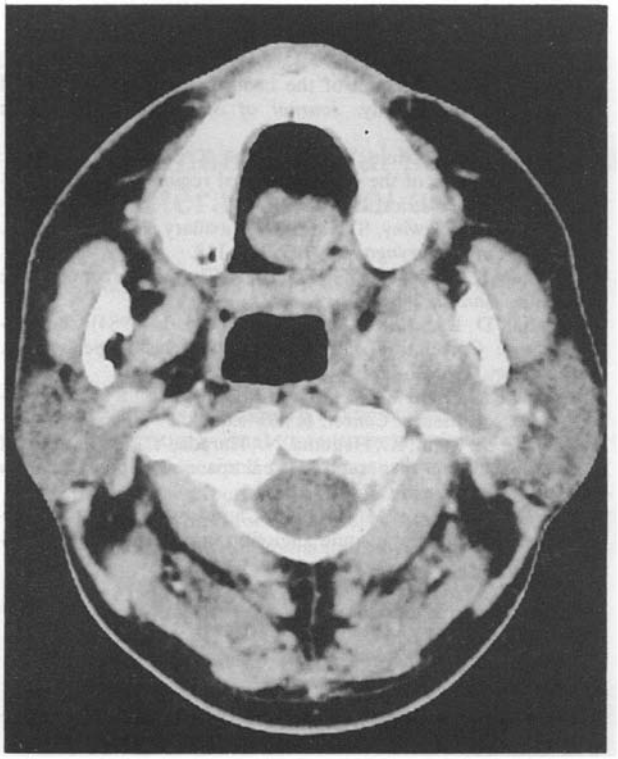

FIG. 1
CT scan showing tumour in the left parapharyngeal space.

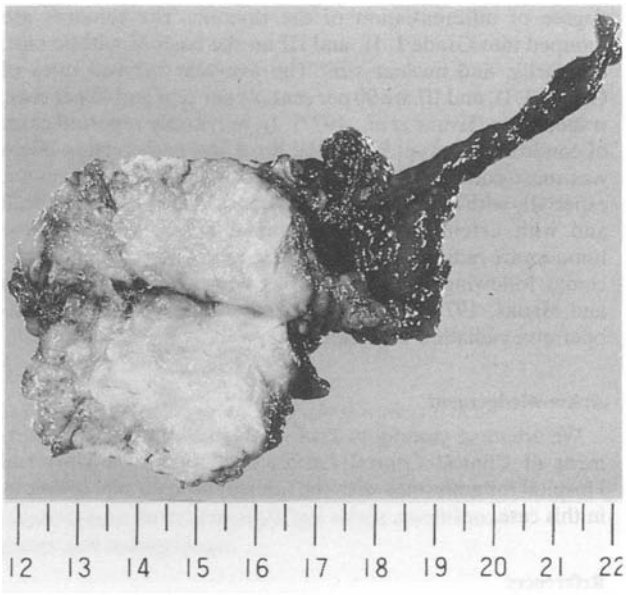

FIG. 2
Cut section of gross specimen.

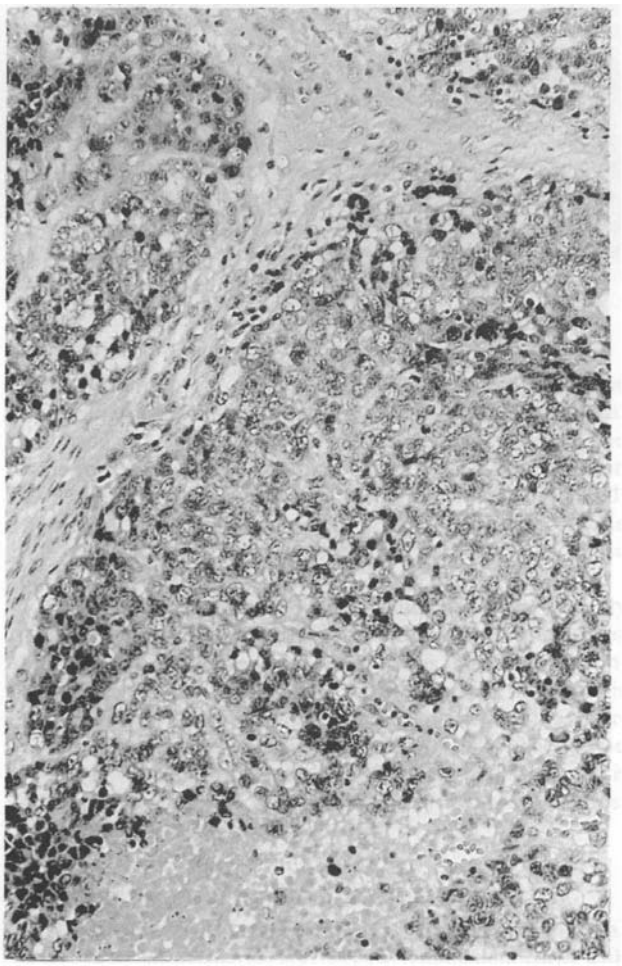

FIG. 3
Chondromatous nodules with numerous chondroblasts. Haematoxylin and eosin ×200.

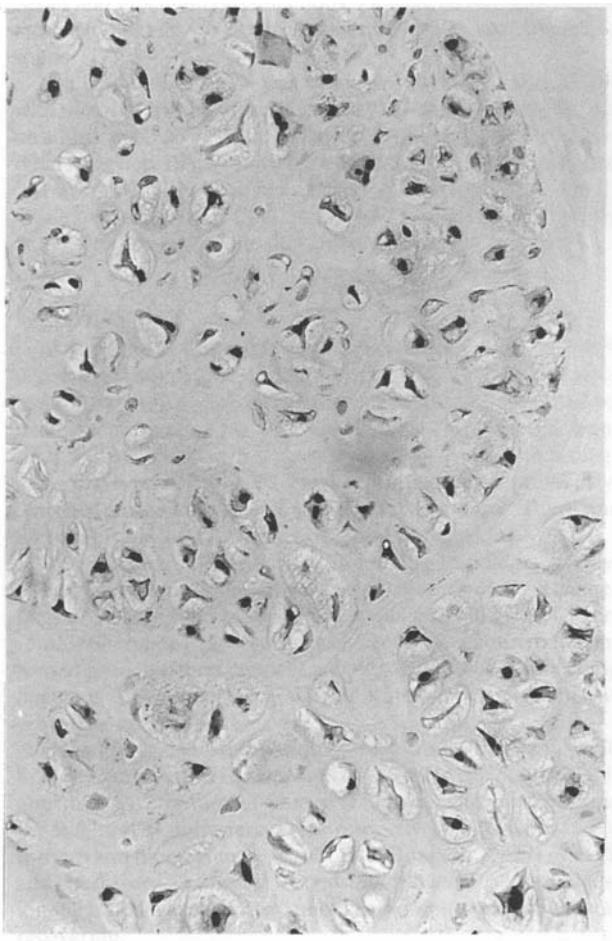

FIG. 4
Chondrosarcoma with cellular zones, large irregular nuclei and multinucleated cells. Haematoxylin and eosin ×100.

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degree of differentiation of the tumour. The tumours are grouped into Grade I, II, and III on the basis of mitotic rate, cellularity, and nuclear size. The five-year survival rates of Grades I, II, and III are 90 per cent, 81 per cent and 43 per cent, respectively (Evans *et al.*, 1977). In previously reported cases of condrosarcoma arising in the head and neck region, death was most commonly caused by local recurrence of tumour, especially with involvement of the bones at the base of the skull and with extension into the cranial cavity. Usually these tumours are radioresistant; however, regression of chondrosarcomas following radiotherapy has been reported (Peddison and Hanks, 1971). In our case, external excision and postoperative radiation was effective.

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