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Pathology in Focus

Chondromyxoid fibroma of the nasal bone with extension into the frontal and ethmoidal sinuses

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Abstract
Chondromyxoid fibroma is a rare benign tumour whose histological appearance may easily be misinterpreted as chondrosarcoma. It has a tendency to recur locally unless completely excised. A rare case of the tumour affecting the nasal bone with extension into the frontal and ethmoidal sinuses and impingement on the cribiform plate is presented. Complete excision was achieved by the craniofacial resection approach.

Key words: Fibroma, chondromyxoid; Nasal bone

Introduction

Chondromyxoid fibroma (CMF) is a rare bone tumour which was first described by Jaffe and Lichtenstein (1948) when they discovered three cases of this tumour while reviewing their chondrosarcoma files. They stressed the importance of the tumour as a distinctive benign tumour likely to be mistaken for chondrosarcoma especially. The World Health Organization (WHO) defines CMF as 'a benign tumour characterized by lobulated areas of spindled-shaped or stellate cells with abundant myxoid or chondroid intercellular material, separated by zones of more cellular tissue rich in spindled-shaped, or rounded cells, with varying numbers of multinucleated giant cells of different sizes; large pleomorphic cells may be present and can result in confusion with chondrosarcoma' (Schajowicz, 1981).

CMF has a predilection to occur in the long bones, with lesions of the femur and tibia accounting for half of all cases and lesions of other bones of the extremities accounting for most of the rest (Wilson *et al.*, 1991). CMF of the cranial and facial bones is very rare and most reported cases had occurred in the mandible (Grotepass *et al.*, 1976; Browne and Rivas, 1977; Pinholt *et al.*, 1986; Lustmann *et al.*, 1986; Muller *et al.*, 1992; Lingen *et al.*, 1993) and the maxilla (Damm *et al.*, 1985; Lustmann *et al.*, 1986; Fujii and Eliseo, 1988). Cases have also been described in the mastoid bone (Kitamura *et al.*, 1989), pterygopalatine space (Toremalm *et al.*, 1976), zygoma (Carr *et al.*, 1992) and petrous sphenoid junction (Frank *et al.*, 1987).

We present a case of CMF arising from the nasal bone with extension into the frontal and the ethmoidal sinuses. We believe the tumour in this unusual site has not previously been described in the literature.

Case report

A healthy 57-year-old Caucasian woman presented with a two-year history of a slowly expanding, painless, swelling over the bridge of the nose. Her only complaint was that her sunglasses would no longer fit properly. There was no history of nasal obstruction, headaches or visual disturbances. Clinical examination revealed broadening of the nasal bridge due to a smooth, firm, nontender swelling (Figure 1). There was no clinical evidence of intranasal extension and the rest of the physical examination was unremarkable. The initial clinical impression was that the swelling represented a midline dermoid.

Cranial CT scan (Figure 2) demonstrated a mass arising at the nasal bridge extending posteriorly into the frontal sinus and eroding into the superomedial part of the right orbit. An MRI scan (Figure 3) was also performed which excluded any intracranial extension.

An excisional biopsy of the lesion was performed via an osteoplastic frontal approach. The tumour was noted to have a lobulated appearance and stripped very easily out of the frontal sinus which showed no sign of bony erosion. The orbital periosteum, as noted on the CT scan, was exposed in the medial part of the orbit. A lot of abnormal granular tissue was found around the tumour, extending especially into the right ethmoidal cells and into the left of the nasal bridge. These tissues were carefully dissected out but, due to the surgical approach employed, it was not possible to safely achieve complete excision of all the granular tissue.

The main tumour bulk consisted of a mass of firm brown tissue, measuring 2.5 × 2.0 × 1.5 cm. Microscopy revealed a lobulated tumour lying beneath the respiratory mucosa (Figure 4). The tumour was composed of bands and lobular collections of partly chondroid, partly fibromyxoid tissue containing numerous round and spindle-shaped cells showing little cellular or nuclear pleomorphism and no mitotic activity (Figures 4, 5 and 6). Focal areas of cystic degeneration and cartilage differentiation were present within the tumour. The tumour matrix showed a metachromatic reaction with alcian blue and toluidine blue stains. Lobules and bands of tumour were well circumscribed, and a few contained multinucleated giant cells at their edges (Figures 5 and 6). The tumour also showed a well-defined border where it abutted bone and did not infiltrate between bone trabeculae. Tumour cells were positive for S100 protein and vimentin

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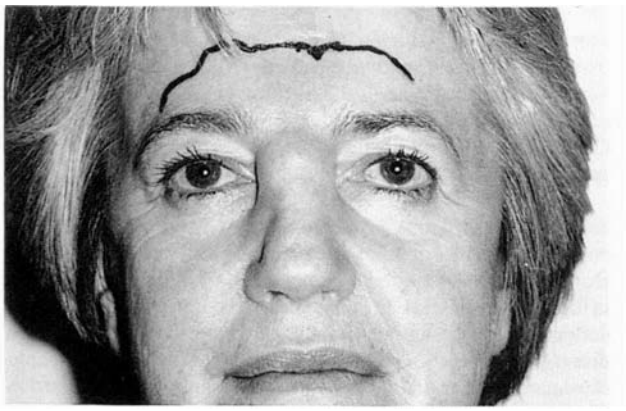


FIG. 1

Photograph of the patient demonstrating the nasal bridge swelling. The markings on the forehead map the outline of the frontal sinus and was used as an aid to the osteoplastic flap procedure.

but negative for epithelial membrane antigen and cytokeratin intermediate filaments, thus distinguishing this lesion from an epithelial tumour or chordoma. Amongst other morphological features, the lack of cellular and nuclear pleomorphism or mitotic activity and the absence of bone destruction associated with tumour infiltration did not favour the diagnosis of mesenchymal chondrosarcoma (a spindle cell tumour showing focal areas of cartilage differentiation) The appearance was considered to be most in keeping with that of a chondromyxoid fibroma, a diagnosis which was confirmed on referral.

A further CT scan was performed which demonstrated residual lesions in the nasal bridge, the anterior ethmoidal cells, and in the region of the cribiform plate. A craniofacial resection of the residual tumour was performed removing the ethmoid sinus, the anterior part of the sphenoid sinus, the medial wall of the right orbit, the whole of the upper nasal skeleton and glabellar skeleton and frontal sinus en bloc. Reconstruction of the glabellar area and the nasal bridge was achieved using bone grafts supported by plates. Further combination of bone grafts was used to reconstruct the floor of the anterior cranial fossa centrally and the medial walls of both orbits. Medial canthopexy was carried out through these bone grafts transnasally and further bone chips laid on the anterior cranial fossa in the orbital roof areas on both sides. Two galeal frontalis flaps were raised, one on each side of the midline. One was passed deep to the nasal bone reconstruction to line the upper part of the nasal cavity and another was passed above the bone graft reconstruction of the anterior cranial fossa to provide a vascular layer there.

The patient had remained well six months after the surgery and is being followed-up on a regular basis.

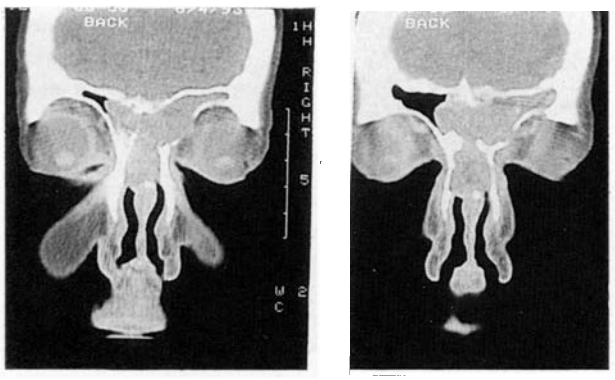
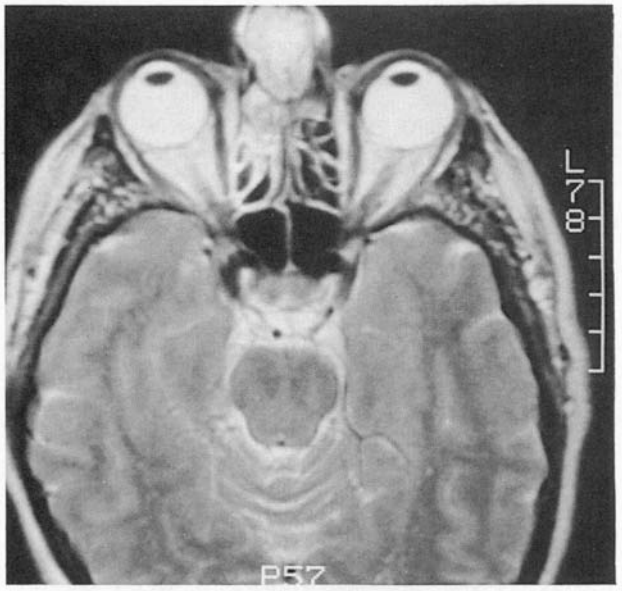
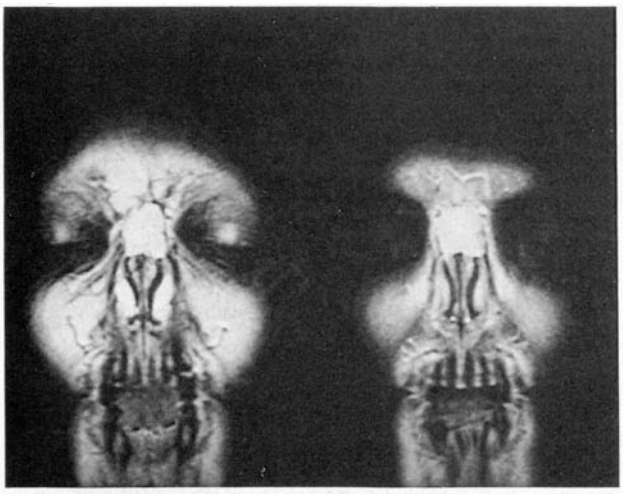


FIG. 2

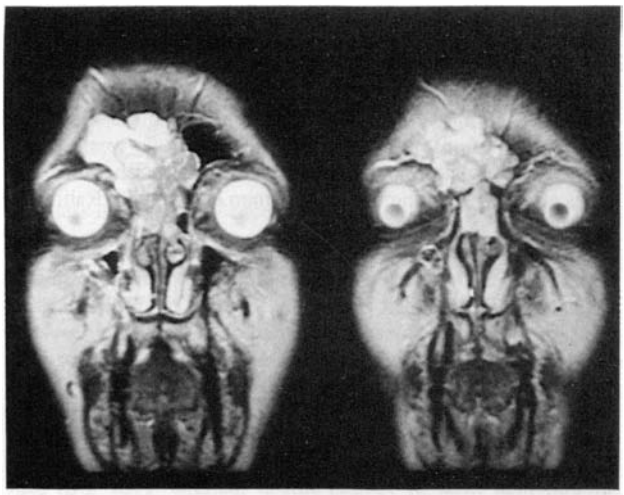
CT scans demonstrating the tumour extending from the nasal bridge into the frontal sinus.



(a)



(b)



(c)

FIG. 3 (a–c)

MRI scans demonstrating extension of the tumour from the nasal bridge into the ethmoidal and frontal sinuses.

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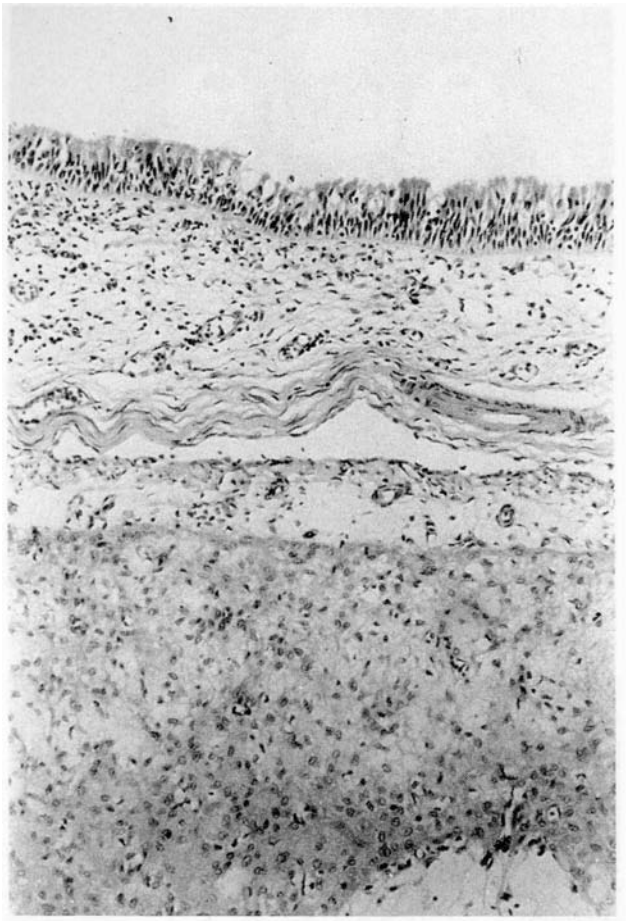

FIG. 4

Low power view of tumour lying beneath respiratory mucosa. (H&E; ×40).

Discussion

A recent collective review by Wilson *et al.* (1991) of data from their own series and eight previously published major series on up to 365 cases of CMF revealed that patients with CMF are usually young, with most (75 per cent) presenting in the first three decades of life, and a peak incidence in the second decade of life. Cases have also been reported in patients as young as three years (Zillmer and Dorfman, 1989) and as old as 79 years (Feldman *et al.*, 1970).

Pain and swelling over the affected areas are the commonest symptoms although some may present as asymptomatic incidental X-ray findings, notably in tumours affecting the ilium and ribs. The duration of symptoms ranges from two weeks to 20 years. Pathological fractures have also been described in some cases affecting the long bones (Feldman *et al.*, 1970; Rahimi *et al.*, 1972).

CMF has a characteristic but nonspecific radiographical appearance and may often mimic more common tumours. Feldman *et al.* (1970) in their review of 207 cases of CMF described the typical radiographical appearance of the lesion in the long bone as a circumscribed, eccentrically situated, round or oval metaphyseal defect with its long axis directed along the long axis of the bone. Wilson *et al.*(1991) pointed out that the problems with the radiological diagnosis of CMF results more from the diversity of sites of involvement and the relatively rarity of the tumour than from its radiographical features. They also suggested that CMF should always be considered as a diagnostic possibility when evaluating solitary bone lesion with geographical destruction that has a lobulated margin, septation, cortical expansion and/or sclerotic rim. The presence of two or more of these features in a patient who is in the second or third decade of life sharply increases the diagnostic possibility of CMF. However, in the rare situation where the tumour is mainly extraosseous, as in the case of our patient, radiological findings may have little to contribute to the histological diagnosis. Lingen *et al.* (1993) reported a similar case of extraosseous mandibular CMF which was initially misdiagnosed as a peripheral giant cell granuloma.

More than 45 years after Jaffe and Lichtenstein (1948) described the difficulty in differentiating CMF from chondrosarcoma, the diagnostic challenge still remains real. Zillmer and Dorfman (1989) reported a 22 per cent initial misdiagnosis rate in the 36 cases of CMF in their series and pointed out the possible danger of major amputation being performed for a benign disease. Conversely, Gallia *et al.* (1980) reported a case of chondrosarcoma of the mandible which was initially misdiagnosed as CMF and resulted in inadequate initial treatment and subsequent recurrence.

The two most common modes of treatment for CMF are either en bloc excision or thorough currettage. The latter was advocated by Jaffe and Lichtenstein (1948) in their original paper. Rahimi *et al.* (1972) advised en bloc excision as the treatment of choice due to the relatively high rate of recurrence after curettage alone. Another argument for radical excision of the tumour was the possibility of malignant transformation although the risk is extremely low unless radiotherapy is employed. Feldman *et al.* (1970) studied 26 cases of recurrence and concluded that at least 17 cases could be attributed to incomplete tumour removal.

Radical removal of long bone CMF usually poses little difficulties. However, wide excision of tumour involving the craniofacial skeleton may result in severe functional and cosmetic morbidity. Lingen *et al.* (1993) reviewed 18 cases of CMF of the

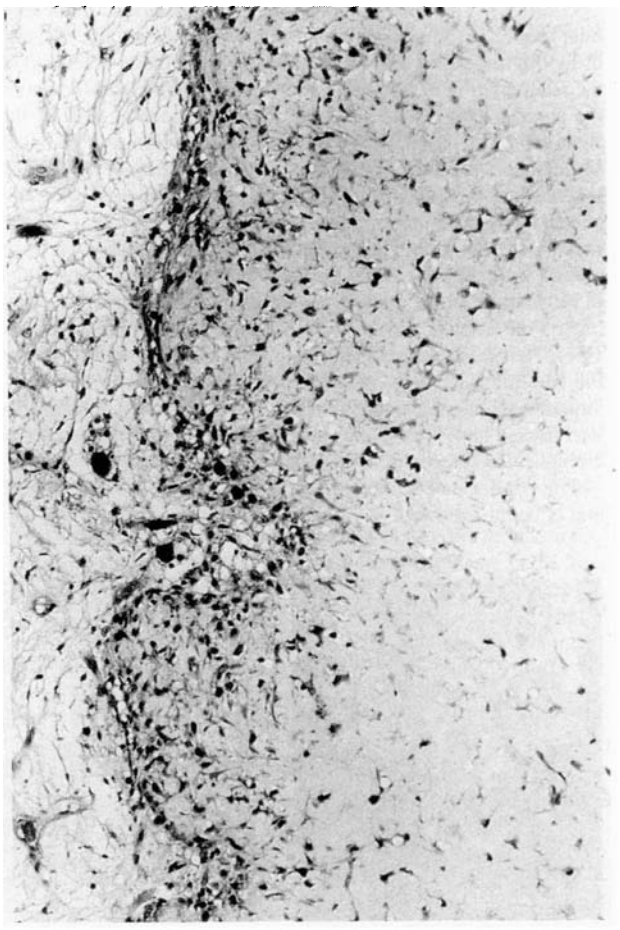

FIG. 5

Rounded, well-defined edge of expanding tumour composed of round and spindled-shaped cells in chondromyxoid matrix. (H&E; ×250).

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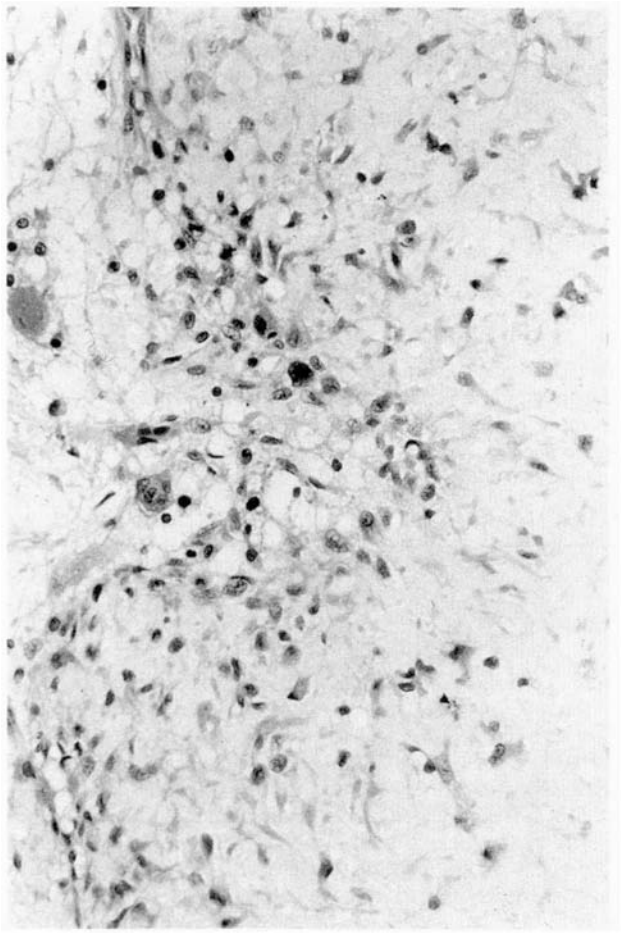

FIG. 6

High power view of tumour showing absence of cellular and nuclear pleomorphism in tumour cells and giant cell at edge of tumour lobule. (H&E; ×400).

jaw, 12 of which were treated either by curettage or enucleation and found two cases of recurrence. Most authors now recommend thorough curettage of the tumour followed by careful periodic surveillance for recurrence in cases of craniofacial CMF. In our patient, excision after the initial osteoplastic frontal procedure was felt to be inadequate in controlling the disease because of histological and radiological evidence of residual tumour. En bloc excision was made possible through a craniofacial resection approach. Complete excision of the tumour should help to minimize the chances of a local recurrence of the disease which may become surgically more inaccessible and less curable.

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