CASE REPORTS

Dedifferentiated chondrosarcoma arising in the orbit

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Figure 1 Chondrosarcoma limited to the right orbit, causing inferolateral ocular displacement, proptosis, and a minor degree of medial ptosis.

Figure 2a

Figure 2b

Figure 2 Axial CT scan (a) shows a well-defined uniform soft tissue mass on the medial orbital wall, with no evidence of sinus involvement. Superior extension of the tumour is demonstrated on coronal scan (b).

Abstract

Orbital chondrosarcomas are extremely rare and are usually an extension of tumours involving the paranasal sinuses. A unique case of dedifferentiated chondrosarcoma arising solely within the orbit is presented.

Most chondrosarcomas arise in the axial skeleton, while extraskeletal chondrosarcomas frequently affect soft tissues of the extremities and have histologically myxoid features.¹ Chondrosarcomas of the head and neck are uncommon and generally arise from the bones of the nose or paranasal sinuses.¹⁻⁴

We present what we believe to be the first reported case of dedifferentiated chondrosarcoma arising solely within the orbit. Chondrosarcomas confined to the orbit are extremely rare and the previously described examples have been of mesenchymal morphology; the present tumour displayed the unusual feature of histological 'dedifferentiation'.

Case report

A 33-year-old Caucasian male presented in February 1989 with a history of intermittent binocular diplopia; the frequency and severity of diplopia, characterised by vertical and oblique separation of images (especially in supraduction), having increased over 2 years. In June 1989 a superior oblique complete tenotomy was performed to relieve an abnormal head posture associated with a right-sided Brown's syndrome. Visual function at that time was recorded as normal.

Right proptosis was evident by November 1989 and the patient was referred to the Orbital Clinic at this hospital.

At referral the patient had normal visual function in both eyes. The right globe was displaced inferiorly and laterally (by 4 mm and 7 mm, respectively) and showed 7 mm of relative proptosis. All movements of the right globe were reduced, especially that of supra-laeovoduction. A mass was present superiorly and medially in the right orbit, causing a minor ptosis medially with normal levator function (Fig 1). Periorbital trigeminal nerve function, the corneal reflexes and the findings on systemic examination were normal, and there was no cervical lymphadenopathy. The patient's general health was good, and there was no clinical evidence of multiple enchondromatosis (Ollier's disease).

Orbital computed tomography (CT) scans showed a soft tissue mass of uniform radio-

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lucency, situated superomedially in the right orbit and displacing, rather than infiltrating, the normal intraorbital structures (Fig 2). The well defined mass was closely applied to (and possibly arising from) the medial orbital wall, which was bowed medially; the paranasal sinuses were otherwise normal.

Anterior (external) ethmoidectomy was performed in April 1990 to provide adequate extra-periosteal access to the medial wall of the right orbit. The periosteum of the medial orbital wall was intact, without breach by extraorbital extension of the tumour. A bilobed purple, friable tumour was resected which, although separate from orbital structures, did not have a defined capsule.

There has been no evidence of tumour recurrence by 1 year after surgery.

Pathology

The tumour comprised two distinct histological

patterns. Features of a cellular cartilaginous tumour were present, with cellular pleomorphism, hyperchromasia, and binucleate cells, these being features of a low-grade chondrosarcoma (Fig 3a). A highly cellular spindle cell sarcoma was evident in other parts of the tissue, with cells arranged in bundles and whorls, and with a storiform pattern in places. Mitoses were frequent in this high grade sarcoma, the tumour resembling a malignant fibrous histiocytoma (Fig 3b).

The occurrence of a malignant fibrous histiocytoma in the presence of a low grade chondrosarcoma is the typical pattern for 'dedifferentiated' chondrosarcoma.

Discussion

Chondrosarcomas outside the axial skeleton are rare and tend to occur in the head and neck. Orbital involvement is extremely rare¹⁻⁴ and is often associated with tumours affecting, or perhaps originating in, the nose or paranasal sinuses.

The tumour in the present case produced symptoms for at least 2 years before resection, although the clinical rate of tumour progression increased markedly in the 6 months before surgery. Acceleration of tumour growth, with a worsening of clinical prognosis, is commonly observed with chondrosarcomas that undergo 'dedifferentiation'.

A 'dedifferentiated' chondrosarcoma is characterised by the presence of a poorly differentiated sarcoma (Fig 3b) in an otherwise typical low grade chondrosarcoma (Fig 3a). Accelerated tumour growth is probably due to this 'dedifferentiation' of the parent tumour, with formation of a more aggressive, highly mitotic, and poorly differentiated neoplasm. The latter tumour is generally undifferentiated sarcoma, fibrosarcoma, osteosarcoma, rhabdomyosarcoma, or (as in the present case) a malignant fibrous histiocytoma.⁵⁻⁷

The prognosis for patients with chondrosarcomas is dependent on histopathological grading. Patients with low grade tumours in bone have a 78% survival at 5 years, but with tumours of moderate or higher grades the survival decreases to 53% and 22%, respectively.⁸ Recurrent tumours tend to a higher, more aggressive, grade, and wide surgical excision provides the most effective primary therapy. Most chondrosarcomas respond poorly to radiotherapy or chemotherapy.

There are a number of histopathological variants of chondrosarcoma: clear cell chondrosarcomas (usually arising in bone) and myxoid-chondrosarcomas (often in the extremities) both have a better prognosis than other variants. Mesenchymal and dedifferentiated chondrosarcomas have a worse prognosis, and many therapeutic combinations have been tried for these tumours.

In the head and neck the wide excision of these tumours (the treatment of choice) is often limited by vital structures; this probably contributes to a worse prognosis for tumours in these locations. Although resection in the present case appeared to be complete, treatment of tumour recurrence

Figure 3a

Figure 3b

Figure 3 Histopathology of dedifferentiated chondrosarcoma: sections showing features of (a) low grade chondrosarcoma and (b) high grade malignant fibrous histiocytoma. (Haematoxylin and eosin: (a) $\times 122$, (b) $\times 60$.)

would be difficult. Secondary exenteration would be complicated by the previous transtethmoidal resection, and the response to radiotherapy is relatively poor with high grade chondrosarcomas.

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FIFTY YEARS AGO

Eye injuries among stone breakers

Among the writer's very early recollections of his childhood when he lived in the depth of the country are the village blacksmith in his smithy, the pedlar and his pack, a morose individual, the local butcher, who still wore the agriculturalists' smock frock, and the road mender breaking stones at the side of the road. The last named wore large protective goggles to avoid eye injuries. He was a formidable figure to the infant mind, and the goggles, made we believe of a fine wire mesh, added to the feeling of fear which his appearance at first sight engendered. In reality he was a most respectable member of my father's congregation and, without the goggles, very pleasant company.

We have recently been making enquiries as to the frequency of eye injuries among the flint knappers of East Anglia. This must certainly be one of the oldest occupations in the world and we are informed that injury to the eyes is exceedingly rare among them. Much of their work is done on a leather shield strapped

on to the left leg above the knee, though the finer work is done on a bench. The knapper knows by long experience how to place his flint. These large flints mostly have flaws in them and the knapper percusses with his hammer with light taps until he has found the right spot and then gives a smart blow on it. Fragments appear to fly downwards and sideways and perhaps this may account for the rarity of eye injuries. This part of his work is rather different from that of the road mender whose fragment of stone lies on a bed of similar fragments, or on the ground nearby, with very little resilience beneath it. The bench work would seem more nearly to approximate that of the road mender, but so far as we could learn injury to the eye is very rare and the few knappers who still ply their trade do not wear, so far as we know, any form of eye protection.

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