CASE REPORTS

Monostotic fibrous dysplasia of the orbit: an unusual lacrimal fossa mass

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Abstract

A case of monostotic fibrous dysplasia involving the orbit, presenting with a short history of painful, progressive proptosis and a lacrimal fossa mass is reported. There were no clinical or radiological clues to the diagnosis which was made on frozen section at the time of open surgical biopsy. The features of orbital fibrous dysplasia are reviewed and a possible mechanism for this rare clinical presentation of fibrous dysplasia is discussed.

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Orbital involvement by fibrous dysplasia is well recognised and may present clinically in several ways, including proptosis. This case report details a rare presentation consisting of a short history of painful proptosis associated with globe displacement and an apparent lacrimal fossa mass.

Case report

A 41-year-old Egyptian female presented with a 4 month history of progressive diplopia, reduction of vision in the left eye, and increasing pain in the upper temporal quadrant of the left orbit where a firm tender mass was palpable. There was no history of trauma. The patient was otherwise in good health with no history of significant medical or surgical illness. She was taking no medication. A careful review of systems was non contributory. Her most recent pregnancy was 5 years ago.

On examination the right eye and adnexae were normal. The left visual acuity was reduced to 6/24. There was a fullness in the upper temporal quadrant of the orbit and a firm, slightly tender mass was palpable in the lacrimal

Figure 2 Coronal CT scan showing large mass in the position of the lacrimal gland. Note the calcification in the mass.

fossa region. The globe was proptosed 7 mm axially and 2 mm inferiorly (Fig 1). There was a significant reduction of elevation and abduction of the left eye. Examination of the globe was normal. General physical examination was normal; in particular, there was no clinical evidence of thyroid disease, nor any abnormal skin pigmentation or bony deformity.

Coronal and axial computed tomographic (CT) scanning of the orbits was performed and revealed an extraconal mass lesion which involved the superior and lateral aspects of the left orbit and contained some calcification (Fig 2). The lesion was smoothly irregular in outline and depressed the globe. There was evidence of early cortical bone erosion by the lesion in the left upper lateral orbital margin and the frontal process of the zygomatic bone. The lacrimal gland could not be seen separately from the mass. Full blood count, ESR, biochemical profile, liver function tests, and thyroid function tests were normal except for the serum alkaline phosphatase which was elevated at 284 units/l (normal <160 units/l).

The history and clinical findings were considered consistent with a diagnosis of lacrimal gland carcinoma and therefore a trans-septal orbital biopsy was performed. At operation a bulky mass was found in the position of the lacrimal gland.

Figure 1 Frontal view of patient. Note the inferior globe displacement.

Figure 3 Operative view showing the tumour mass and its origin from the frontal bone.

Its surface was thick and pale but on cutting it proved to be gritty, friable, and haemorrhagic. Histopathological examination revealed a fibro-osseous lesion containing metaplastic bone with irregular birefringence consistent with fibrous dysplasia. About one third of the tissue removed was fibrous with foci of multinucleated giant cells

and prominent recent haemorrhage, features found in giant cell reparative granuloma.

Five days later a modified Wright's lateral orbitotomy was performed. At surgery the mass was found to be surrounded by a capsule and superficial to the periorbital. The mass appeared to arise from bone and it was not possible to dissect the mass free from bone (Fig 3). An osteotome was therefore used to remove the mass and curettage of the orbital bones was then performed to remove any residual tumour. Histopathological examination confirmed the diagnosis of fibrous dysplasia with areas of giant cell (reparative) granuloma (Figs 4 and 5). The latter included acute haemorrhage, fibrocytes, and multinucleated giant cells.

Following surgery, the patient's visual acuity returned to normal and the other physical findings resolved. When last reviewed some 6 months postoperatively, orbital and ocular examination were normal apart from the surgical scars.

Comment

Fibrous dysplasia is an uncommon benign bony disease of unknown aetiology which typically presents in children or young adults. First recognised in 1937 by Albright as a syndrome characterised by 'osteitis fibrosa disseminata', areas of pigmentation, and endocrine dysfunction, it later became clear that the bony lesions could develop in the absence of the other features of Albright's syndrome and that the disease could be monostotic as well as polyostotic in distribution.¹ The disease is not heritable and is seen with equal frequency in both sexes.

Monostotic involvement usually involves the craniofacial bones or ribs with the sphenoid and frontal bones being the most commonly affected. Polyostotic lesions may involve any bone and be widespread but typically the lower limbs are predominantly affected. Up to 50% of girls with the polyostotic form have associated macules of abnormal skin hyperpigmentation (Coast of Maine lesions) and precocious puberty. Abnormal skin pigmentation may also be seen in boys.¹

Orbital involvement is common as the sphenoid and frontal bones are sites of predilection for the disease. The typical patient presents in the first three decades of life with slowly progressive painless proptosis variably associated with non-axial globe displacement, dystopia, and facial disfigurement. It is frequently progressive but is thought to be self limiting with a variable end point in the second or third decades of life.²

In contrast, our patient was aged 41 years and presented with a short history of rapidly increasing proptosis, a mass lesion, globe displacement, and pain typical of an infiltrative process. The radiological findings were consistent with a mass lesion in the lacrimal fossa associated with bone destruction. The patient had no clinical evidence of bone disease, there was no history of precocious puberty, and there were no skin lesions. Indeed until the trans-septal biopsy was performed the working diagnosis was lacrimal gland carcinoma. Although it is well recognised that fibrous dysplasia produces proptosis, it rarely presents with the history seen in our patient. A

Figure 4 The areas of fibrous dysplasia with metaplastic bone trabeculae merge with non-bony areas showing acute haemorrhage, fibrous tissue, and multinucleated giant cells. At the edge of the lesion the cortical bone was generally very thin or absent (magnification $\times 8$).

Figure 5 A higher power view showing two metaplastic bone trabeculae and adjacent fibrohaemorrhagic tissue with giant cells consistent with giant cell reparative granuloma (magnification $\times 20$).

review of the literature was able to identify only one similar reported case where proptosis evolved over 1 month; however this was associated with dysaesthesia but no pain.³

The histopathology of the mass removed at orbitotomy revealed the features of both fibrous dysplasia and giant cell reparative granuloma. Giant cell reparative granuloma consists histologically of haemorrhagic masses with a spindle cell stroma containing osteoblastic giant cells and haemosiderin.² Giant cell reparative granuloma is a benign process, which occurs in association with trauma and intraosseous haemorrhage; it is part of the spectrum of reactive bone conditions commonly seen in a number of bone diseases including fibrous dysplasia.

The haemorrhage and reparative granuloma

may well explain the most unusual clinical presentation of this patient. It is likely that this patient had a small localised area of monostotic fibrous dysplasia in the zygomatic process of the frontal bone which was asymptomatic. Bleeding then occurred in an area of the fibrous dysplasia either spontaneously or following trauma. The resultant swelling and reparative granuloma was then responsible for the rapidly evolving painful proptosis.

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- 2 Rootman J. *Diseases of the orbit*. Philadelphia: Lippincott, 1988: chapter 12.
- 3 Retter RH. Unilateral proptosis due to monostotic fibrous dysplasia. *Ann Ophthalmol* 1976; 8: 45-9.