Primary orbital liposarcoma: clinical and computed tomographic features

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Abstract
Liposarcoma of the orbit is rare. The clinical and radiological characteristics of two cases, of myxoid and pleomorphic types, are reported. In neither case was the histological diagnosis evident before surgery.

Primary orbital liposarcoma is extremely rare. Sixteen cases, mostly myxoid and none definitely identified as pleomorphic, have been reported in detail in the literature. The two patients described here are the fourth and fifth to be seen in the Orbital Clinic at Moorfields Eye Hospital since 1982. The preceding patients, all of whom had myxoid tumours, have been reported on previously. To our knowledge the clinical and radiological features of pleomorphic liposarcoma arising in the orbit are described for the first time.

Case reports

CASE 1
A 69-year-old woman complained of progressive right proptosis over six months. She had vertical diplopia and difficulty in seeing colours clearly with the right eye. Her general health was excellent.

Her best corrected vision was 6/9 in the right eye and 6/6 left. She made four errors on the Ishihara test plates with the right eye, and was thought to have an afferent pupillary defect. The globe was displaced 9 mm forwards and 4 mm downwards. Ocular motility was restricted, particularly in elevation. A non-tender mass was palpable in the upper orbit. General examination was normal, as was a chest radiograph.

Computed tomography (CT) of the orbits was obtained in sagittal and coronal planes after intravenous injection of iodinated contrast medium (Fig 1). A well defined, rounded intracranial mass of low density was shown, lying just above the globe.

The mass was explored via an upper lid skin incision, the levator palpebrae superioris and superior rectus muscles being retracted medially. Anteriorly the tumour was well circumscribed, but posteriorly it appeared to blend with the orbital fat. It was pale, yellow-grey, and petechial haemorrhages appeared on its surface as it was touched. The incisional biopsy specimen floated in 10% formaldehyde solution.

Histological examination revealed a tumour with numerous rounded spaces which on frozen section contained lipid. Some cells were spindle shaped and lay in a mucinous matrix, while others showed a signet ring form. There was nuclear hyperchromatism, scattered cells having enlarged, pleomorphic nuclei; some of the nuclei were vesicular. Numerous capillaries were present throughout the mass. The appearances were typical of a myxoid liposarcoma (Fig 2).

Over the next four weeks the proptosis increased by 2 mm. Orbital exenteration was performed, with removal of the upper lid and adequate clearance beyond the posterior margin of the tumour. There has been no evidence of recurrence for more than a year since surgery.
Figure 2: Case 1. Myxoid liposarcoma with nuclear hyperchromatism and occasional signet ring cells. The numerous clear spaces contained lipid on frozen section. (Haematoxylin and eosin, ×225.)

Figure 3: Case 2. CT without intravenous contrast medium. Direct axial (A) and reformatted oblique parasagittal (B) images: a well defined, lobulated mass over 2 cm in diameter lies within the muscle cone, superomedial to the optic nerve, slightly flattening the globe. The mass is generally isodense with brain; its less dense posterior portion is still denser than the vitreous. The roof of the orbit is very thin, possibly eroded.

Figure 4: Case 2. Pleomorphic liposarcoma. The nuclei are variable in size, many having large, prominent nucleoli. The clear spaces also contained lipid on frozen section. (Haematoxylin and eosin preparation, ×225.)

CASE 2

A 52-year-old man noted blurred vision in the right eye seven months before his presentation to the orbital clinic, followed two months later by proptosis. His vision corrected to 6/9, but he had mild disc swelling and 3 mm of proptosis. This increased to 5 mm over 3 months, and he developed diplopia on left lateral gaze. He had no pain.

In the orbital clinic his best corrected acuity was 6/24 in the right eye and 6/5 left. He read all the Ishihara plates, but was slower on the right. There was a mild right relative afferent pupil defect. The globe was displaced 5 mm forwards and inferolaterally, with limited elevation and adduction. The optic disc was grossly swollen, with engorged retinal veins, but there were no choroidal folds. The upper lid was full. General examination and chest radiograph were normal.

Plain films of the orbits also showed nothing abnormal, but CT showed a well defined mass within the muscle cone, containing areas of lower density within it (Fig 3).

At lateral orbitotomy a large, purplish, lobulated tumour was found intraconally, abutting the globe, the medial rectus muscle, and the upper surface of the optic nerve. It appeared to be well encapsulated, and was dissected free of the surrounding structures. A large artery entering its posterior pole was divided and the mass was removed in one piece.

Histological examination (Fig 4) disclosed a liposarcoma of extremely variable morphology, with numerous thin walled vessels, lipoblasts, and large, deeply eosinophilic giant cells. Mitotic figures were infrequent, but foci of inflammation and haemorrhage were present. There was no true capsule. The histological diagnosis was therefore pleomorphic liposarcoma.

The orbit was exenterated, together with the scar, but the lids were spared. Examination of the excised tissue showed a single nodule of tumour, less than 1 mm in diameter, well clear of the resection margin. One year later there was no evidence of local recurrence or distant metastasis.

Discussion

Because of its rarity liposarcoma does not usually enter into the differential diagnosis in patients with orbital masses. The 16 primary orbital liposarcomas reported in detail1-12 affected men and women equally, their ages ranging from 5 to 77 years, mean 34.

Reports of the results of imaging are sketchy. Plain radiography commonly shows nothing abnormal, but it may show enlargement of the orbit, sometimes considerable, with thinning and inferior displacement of the floor.1 Echo-}


graphy may indicate a relatively well defined, multiseptate lesion with non-specific areas of high echogenicity.13 In the few patients who have undergone CT, none of whom had pleomorphic tumours, findings have varied from well defined, homogeneous lesions of soft tissue density, not
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