Liposarcoma of the orbit: a clinicopathological case report

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SUMMARY Liposarcoma is a very rare primary tumour of the orbit. The present report describes changes seen by computed tomography, carotid angiography, and histopathology and gives a review of the literature.

Liposarcoma is an uncommon mesenchymal tumour. It shows a preference for certain locations such as the lower limbs and the retroperitoneal region.¹ In the orbit only a few cases of histopathologically verified liposarcomas have been reported. It is the object of this report to describe an additional case of primary liposarcoma of the orbit and to give a review of the literature.

Case report

In August 1977 the patient, a 58-year-old woman, was admitted to the University Eye Clinic in Uppsala with painless left-sided exophthalmos of almost one year's duration. Apart from 5 mm protrusion of the left eye she had no clinical abnormalities. Plain x-ray of the orbit was normal. Computed tomography (CT), however, revealed a tumour mass in the mediolateral part of the left orbit (Fig. 1). The attenuation in the tumour varied in different parts, with CT-numbers ranging from +5 to +15 Hounsfield units (HU). Following intravenous contrast injection (60 ml Isopaque Cerebral, Nyco, Oslo, Norway) there was a moderate increase in attenuation, with CT numbers between +20 and +30 HU. By carotid angiography the tumour was seen to be supplied by small branches from the ophthalmic artery, and the tumour displayed a marked blush in the venous phase (Fig. 2a, b). A fine-needle aspiration biopsy contained regular endothelial cells of normal appearance, whereas pathological cell forms were lacking. At surgery a firm and lobulated tumour was easily removed by lateral orbitotomy. The postoperative period was uncom-

plicated. The patient has been followed up for 3½ years and she has not shown any clinical or CT signs of tumour recurrence.

Fig. 1 CT scan showing large retrobulbar tumour in the left orbit and marked exophthalmos.

OPHTHALMIC PATHOLOGY

The tumour measured 33×27×27 mm. It had a firm consistency and a greyish to yellowish cut surface. Histologically the tumour showed both loose and solid areas. In the loose areas the predominant cell type was a rather small spindle to stellate cell with poorly defined boundaries. The nuclei were small and dense, giving an impression of pyknosis. The cells were loosely arranged in a slightly eosinophilic mucoid matrix (Fig. 3a) which stained weakly with alcian blue stain and colloidal iron stain. It was weakly metachromatic with toluidine blue. After digestion with testicular hyaluronidase for 2 hours at 37°C

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Fig. 2a

Fig. 2b

Fig. 2 *Internal carotid angiography. (a) The arterial phase shows delicate tumour vessels (arrows) from the ophthalmic artery. (b) The venous phase shows tumour blush (arrows). The position of the eye globe is seen by the contrast enhancement of the choroid (wedges).*

neither of the stain were positive, thus indicating that the background material consisted mainly of acid mucopolysaccharides. In the periphery of the mucoid areas there was a successive transition into areas with immature lipocytes (Fig. 3b). These cells showed

more definite cytoplasmic borders and their cytoplasm contained coarse vacuoles. The round or oval nuclei showed obvious polymorphism and were placed either centrally or peripherally in the tumour cells. Mitosis or bizarre cell nuclei were not observed.

Fig. 3a

Fig. 3b

Fig. 3 *(a) Myxomatous area of the liposarcoma with stellate and spindle shaped cells. Note the maze of capillaries. (b) Part of the tumour showing lipocytes in different stages of maturation. (HE, $\times 240$).*

In some areas there was a successive change of the cells into mature lipocytes. The solid areas were built up by fibrotic strands crossing through the tumour tissue. The lipocytes stained bright red with scarlet red and black with Sudan black. The lipid drops varied in size from very small to coarse in the immature cells, whereas only large drops filled the mature lipocytes. Throughout the whole tumour there was a rich capillary network. This was most prominent in the areas showing the lowest degree of cellular differentiation. Large vessels were rare. The tumour had a pseudocapsule of condensed tumour cells as its outer limit.

In accordance with the histopathological findings the tumour was diagnosed as a well differentiated myxoid liposarcoma of the orbit.

Discussion

Liposarcomas of the orbit are very rare tumours and to our knowledge only 11 cases have been reported.²⁻¹¹ Various authors^{3,4,12,13} have classified these tumours. According to the classification by Enzinger *et al.*,¹³ which was accepted as the WHO classification of 1969, 4 different types of liposarcomas may be separated, namely, myxoid, embryonal round cell, well differentiated, and pleomorphic. In the present case the histological diagnosis of myxoid liposarcoma was based on the presence of proliferating lipocytes in various stages of differentiation, the myxoid hyaluronidase-sensitive acid mucopolysaccharides, and the prominent capillary network. In the previously reported cases the myxoid type has been predominant.

The examination by CT revealed a retrobulbar tumour with mixed attenuation. The CT characteristics of this kind of tumour located in the orbit has not to our knowledge been described hitherto. The CT characteristics of liposarcoma in other regions such as extremities, mediastinum, and retroperitoneum have been reported.¹⁴⁻¹⁶ In these cases there was a wide range in attenuation values of the tumours (from -90 to +30 HU), and in some cases with low attenuation values the fatty origin of the tumour could be predicted from the CT scan. However, the attenuation in the present tumour did not indicate the lipomatous character of the mass.

Carotid angiography showed a highly vascularised tumour. Several angiographic patterns of liposarcomas have been described,¹⁷⁻²⁰ but they did not differ from those seen in other malignant tumours.

Fine-needle aspiration has been shown to be of great help in diagnosing orbital tumours.²¹ In the present case only endothelial cells were obtained and a definitive diagnosis could not be made. However, in

view of the maze of capillaries in the tumour this finding is not surprising.

Radical surgery is generally regarded as the treatment of choice for liposarcoma. Jones and Jakobiec¹¹ described 2 cases of orbital liposarcoma which both primarily had no other therapy than surgery. In both these cases there were clinical signs of recurrence of the tumour one year later. In the case of liposarcoma reported by Schroeder *et al.*¹⁰ 2 years passed before there were clinical signs of tumour recurrence. In these 3 cases radiation therapy was given after the recurring tumour had been removed by orbital exenteration. In the present case the patient has been followed up for 3½ years postoperatively and she has not shown any signs of tumour recurrence clinically or by CT. According to Schäfer and Wellens¹⁹ radiation therapy should always be given to liposarcomas irrespective of the histological diagnosis. In cases of myxoid liposarcomas irradiation alone may be curative. Cytostatic drugs are recommended only in cases with metastases, where a rate of remission of 40% has been reported.²²

In retrospect it seems that the prognosis of orbital liposarcomas is rather favourable and that the tumour may not recur after orbital exenteration combined with irradiation and finally chemotherapy.

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