LIPOSARCOMA WITH ORBITAL METASTASES*

CASE REPORT

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A WELL-KNOWN anatomical feature of the orbit is its rich fatty content, distributed both inside and outside the muscle cone. The literature contains numerous reports of a variety of orbital tumours, and although angiolipomas and fibrolipomas appear quite frequently, the occurrence of a true lipoma is extremely rare. Strauss (1911) based his diagnosis of primary liposarcoma of the orbit on the predominance of fat in the tumour. In 1960, Enterline, Culbertson, Rochlin, and Brady, in their review of 53 cases of liposarcoma, included one case of metastatic orbital liposarcoma. Fourteen of their cases metastasized, and only two of their patients were still alive at the time of review, and one of these two had metastasis in the lung six years after the original diagnosis. In the other case metastatic liposarcoma of the orbit was diagnosed sixteen years after the original diagnosis of liposarcoma of the thigh had been made. This patient also developed multiple secondaries with eventual invasion of the pelvis. The interesting case here presented is one of primary liposarcoma of the thigh with distant metastases.

Case Report

A man aged 43 years was admitted to the ophthalmic department of Ein Shams University Hospital on July 29, 1963, with a history of three days' right proptosis. The patient first complained of severe right-sided headache together with blurring of vision in the right eye. He had noticed associated progressive right proptosis, and diplopia. He also gave a history of a swelling in the right middle third of the medial side of the left thigh. This swelling had been removed two years prior to admission to hospital, but recurred within a few days of removal.

Examination

Right eye.—Ocular movements in all directions were found to be limited; the fundus showed progressive right papilloedema resulting in total retinal detachment. Although the cornea was covered by tarsorrhaphy, the progressive proptosis led to exposure of the cornea, ulceration, and panophthalmitis.

Left eye.—Normal.

Investigations.—A full blood count showed no abnormality. The Wassermann reaction was negative, the Mantoux test positive. Faeces showed no parasites, ova, or occult blood. Radiographs of the chest showed multiple secondary deposits in both lungs; those of the skull were normal. Angiography, carried out before the onset of hemiplegia, showed no abnormality. The urine was normal.

Progress

Biopsy of the recurrent swelling of the thigh was not carried out, and the pathological report on the original thigh biopsy was not available. The patient developed recurrent attacks of cough, haemoptysis, and left hemiplegia, and died 3 months after admission.

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Post-mortem Findings

Left thigh.—An irregular subcutaneous mass, measuring about $15 \times 10 \times 5$ cm. in diameter, was present in the medial part of the middle third of the left thigh anteriorly; it was soft in consistency and not attached to the deeper structures. On excision, this mass was found to be composed of many nodules which were fixed to each other, but sharply demarcated from the surrounding tissues. The consistency of these nodules was soft and gelatinous; the cut surface was homogeneous and greyish-yellow in colour.

Right eye.—This showed marked proptosis due to the presence of a large soft-tissue mass within the muscle-cone which revealed identical characteristics to those of the thigh lesion.

Lungs.—Both lungs were infiltrated with multiple scattered nodules of similar appearance and consistency to those found in the thigh lesion, and were more dense in the lower two-thirds; the apices were only slightly affected.

Brain.—Weight 1,450 g. Anteriorly, the right temporal lobe was covered by a large haematoma. A small, rounded, soft mass about 3 cm. in diameter, which was easily shelled out, was shallowly embedded within the brain tissue. The cut surface of this mass showed areas of haemorrhage and congestion.

Lymph nodes.—Numerous enlarged para-aortic, inguinal, and mesenteric lymph nodes were seen; they were variable in size and very soft in consistency.

Bones, heart, kidneys, suprarenals, liver, spleen, stomach, and intestines were normal.

Ten per cent. formol saline-fixed specimens from the thigh, orbit, lungs, lymph nodes, and the nodule in the brain were submitted for histopathological examination.

Sections stained with haematoxylin and eosin from the above tissues showed numerous stellate cells, each having an indefinite cytoplasmic boundary with many processes connected with adjacent cells. The nuclei were large and hyperchromatic (myxoid cells). There were also many large round cells containing vacuoles which occupied a major portion of the cytoplasm. The nuclei were large and found on one side, giving the cells a signet-shaped appearance (Figs 1 and 2). There were a few adult fat cells. The stroma between the cells was scanty, with minimal reticulin fibres. Numerous dilated thin-walled blood vessels were present. A few giant cells were seen only in the lung sections (Fig. 3).

FIG. 1.—Section showing signet-shaped cells. Haematoxylin and eosin.

FIG. 2.—High-power view of same section. Haematoxylin and eosin.

FIG. 3.—Giant cells in lung section. Haematoxylin and eosin.

Frozen sections stained with Sudan III showed the signet cells to be filled with fat granules staining deep orange. Sections stained with Mayer's mucicarmine stain showed pink-staining ground substance indicative of mucoid material.

Diagnosis.—Well-differentiated liposarcoma with widespread metastases.

Discussion

Liposarcoma has not hitherto been regarded as a common malignant soft-tissue tumour. It is, however, not so rare as is commonly supposed and can easily escape diagnosis. According to Boyd (1961) it may occur wherever fat is present, but is more commonly found in intramuscular tissue, around joints, and in the retroperitoneal and perirenal regions.

According to Enterline and others (1960) liposarcoma is considered to be a fairly common malignant tumour of soft tissue. Of this group of tumours it is perhaps the most important, since, with correct diagnosis and adequate therapy the condition has an excellent prognosis.

Stout (1944), in his classic paper, divided liposarcoma into four groups: (1) Well-differentiated myxoid type; (2) poorly differentiated myxoid type; (3) round-cell liposarcoma; and (4) mixed liposarcoma.

Enterline and others (1960), following the study of 53 cases of liposarcoma, added a fifth group to the above classification. They called it non-myxoid liposarcoma and described the tumour as being chiefly composed of round cells.

The case presented in this communication agrees histologically with Stout's type I liposarcoma, yet it showed giant cells, found only in the sections taken from the lungs. Enterline and others (1960) were convinced that in all their preparations, type I liposarcoma did not show giant cells, and also that this type of tumour does not give rise to metastases.

Orbital metastases are very rare in cases of liposarcoma. In 1960, Enterline and others reported one such case in which the metastases occurred sixteen years after discovery of the primary which, as in our case, arose in the thigh. It should be noted, however, that in the case here reported, metastases appeared only two years after discovery of the primary.

Summary

A rare, well-differentiated, myxoid type of liposarcoma is presented and discussed. The post-mortem findings and the histological appearances of the tumour as seen in the thigh, lungs, lymph nodes, and orbit are reported.

REFERENCES

- BOYD, W. (1961). "A Textbook of Pathology", 7th ed. Lea and Febiger, Philadelphia.
 ENTERLINE, H. T., CULBERSON, J. D., ROCHLIN, D. B., and BRADY, L. W. (1960). *Cancer*, **13**, 932.
 STOUT, A. P. (1944). *Ann. Surg.*, **119**, 86.
 STRAUSS (1911). *Dtsch. med. Wschr.*, **37**, 239.