OSTEOCLASTOMA OF THE ORBIT*

CASE REPORT

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TUMOURS characterized by the presence of giant cells are not uncommonly seen in sections from orbital masses. The histologic appearance of osteoclastoma as first described by Nélaton (1860) is of tumours consisting essentially of spindle cells, round cells, and multinucleated giant cells with an abundance of nuclei. It is a rare tumour, which until recently had not been differentiated from other fibrous tumours of bone, and usually occurs at the ends of long bones, although a few have been reported in the orbit and in the region of the sphenoid and the labyrinth. This paper reports a case of osteoclastoma of the orbit.

Case Report

A boy aged 5 years was admitted to Ein Shams Hospital complaining of progressive right proptosis. Three weeks before admission he had a raised temperature, which was followed by proptosis and later by profuse watering of the right eye.

Clinical Examination.—On admission, the right eye showed severe, non-pulsating, irreducible proptosis (27 mm. by simple exophthalmometer) and lagophthalmos. Movement was limited in all directions. No mass was detectable around the orbital margins. Ten days after admission the cornea had turned opaque from exposure. Regional lymph nodes were not enlarged.

Investigations.—Radiographs of the orbit were normal. Blood Wassermann was negative and the blood count showed lymphocytosis. Orbitonometry, carried out under general anaesthesia, proved the presence of a solid orbital mass. A clinical diagnosis of a highly malignant tumour was made and exenteration of the orbit was carried out. At operation no bony attachment was revealed and the bones of the orbit were normal.

Pathology

Macroscopic examination of the exenterated orbital tissue showed an intact globe with a firm rubbery mass involving both the inside and the outside of the muscle cone. This mass appeared nodular and well encapsulated. The exenterated tissue was cut vertically through the centre revealing a greyish-white mass with a few areas of haemorrhage. No mass was seen inside the globe, which was slightly compressed by the tumour; the lens was clear and the retina *in situ* (Fig. 1).

Microscopic examination: Sections stained with haematoxylin and eosin show the tumour mass to consist of spindle cells and a large number of giant cells possessing numerous centrally placed nuclei. The cytological picture is fairly uniform with no pleomorphism or active mitosis. Fibrous bands of osteoid tissue extend into the growth. In some areas there is extensive haemorrhage with old blood pigment, and some cystic cavities containing albuminous fluid are present.

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FIG. 1.—Shows the tumour mass surrounding the opened globe which is free from growth.

Posteriorly the tumour appears to be partially encapsulated, especially where it compresses one of the ocular muscles; anteriorly, however, it has developed more invasive properties, and although these tumours are usually benign, malignancy cannot be excluded in this case. The histopathologic picture was typical of osteoclastoma of the orbit (Figs 2 and 3).

Further examination of the sections of the globe showed no abnormal changes except a peripheral corneal vascularization and oedema with perilimbal round-cell infiltration.

The child was discharged from hospital four weeks after the operation when the socket was healthy. Unfortunately, it was impossible to locate him at a later date in order to carry out further radiography to exclude the possibility of von Recklinghausen's disease of bone.

FIG. 2.—General view of the histology showing typical osteoclastoma. Haematoxylin and eosin. $\times 105$.

FIG. 3.—High-power view showing large giant cells with centrally placed nuclei. Haematoxylin and eosin. $\times 435$.

Discussion

This is an exceptionally interesting case, and very few similar cases have been recorded in the literature. Wilson reported a case arising close to the inner margin of the bony orbit, and Azer (1955) published an account of an osteoclastoma arising in close proximity to the lateral orbital wall. The tumour here reported was completely free of the bony orbit, something which makes it not only a great rarity but also a matter of pathological interest. An exhaustive perusal of the literature has failed to reveal any previously published record of an identical orbital case.

The patient in our case was a child of 5 years. In Coley's (1960) extensive review of osteoclastoma, no cases of this condition occurring in patients under the age of 5 years were reported.

Although extraskkeletal bone forms, not infrequently, under a variety of different conditions, the occurrence of true osteogenic tumour outside the skeleton is rare. Fine and Stout (1956) reported 12 cases of osteogenic sarcoma of soft tissues; all tumours showed evidence of osteogenesis, but it was inconspicuous in 6 tumours which were chiefly composed of giant cells and fibrosarcomatous tissue.

According to Boyd (1961), the so-called giant-cell tumours of tendon sheaths and synovial membranes are quite unrelated to the bone tumours, being granulomatous rather than neoplastic in nature.

In a recent paper by Wojnerowicz (1963), a case of giant-cell sarcoma of the mammary gland was reported.

Apart from the osseous system, mammary glands, and neoplasms arising in "myositis ossificans", a giant-cell texture was found in a mesodermal mixed tumour of the uterus, a case recorded by Bickel and Bennett (1954).

The origin of this tumour has been the subject of much speculation and confusion.

There is difference of opinion on practically every aspect of the subject. In 1924 Looser suggested that the giant-cell tumour, classified as osteoclastoma, originated either in regeneration or in inflammatory hyperplasia—a view supported by a number of later workers. More recently, however, Schajowicz (1961) and the majority of subsequent writers agree that the giant-cell tumour is a neoplastic process and that the mononuclear abnormal cells arise from the undifferentiated mesenchymal cell of bone marrow.

The theory of Hanseemann expressed by Nicholson (1918) proposes that when normal cells embark on malignant growth, they demonstrate various stages of dedifferentiation, sometimes to a degree of displaying embryonal characteristics. In support of this theory it has been recognized by Binkley and Stewart (1940) that extraskkeletal osteogenic tumours may develop by a process of metaplasia in various types of tumours.

In view of the absence of any changes in orbital bone in the case here reported, the possibility that this tumour may have arisen from soft tissue must be considered. The presence of orbital inflammation might be regarded as either a predisposing factor or a complication of orbital venous congestion.

Summary

A rare case of proptosis in a child aged 5 years is reported, in which a clinical diagnosis of highly malignant tumour was made.

Microscopic examination, following exenteration of the orbit, revealed an osteoclastoma in which malignant change could not be excluded. The neoplasm is of particular interest in that it was completely unconnected with the orbital bone.

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