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Haemangiopericytoma

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SUMMARY Three cases of orbital haemangiopericytoma are presented. In one the tumour recurred after 22 years and in another after 4 months. Metastasis to the breast, which occurred in the third case, is probably the first to be reported. The management of the cases is discussed. The clinical importance of the haemangiopericytoma lies in its potentially malignant behaviour.

Haemangiopericytoma is a rare vascular tumour. About 50% of cases are malignant.[1] The orbit is an uncommon site,[2] and mostly isolated cases or small series have been reported.[3,4] Distant metastases, though uncommon, occur via the vascular and lymphatic routes, and the lungs are the commonest site for them.[5,6]

We present 3 cases of orbital haemangiopericytoma with marked disparity in their clinical behaviour and in one an uncommon site of metastasis.

CASE 1

A 57-year-old male was admitted to the Rajendra Prasad Centre in August 1979 with the complaint of recurrent proptosis of the right eye for 30 years. At first he had had eccentric proptosis with no visual impairment. He was operated on in 1949 elsewhere and was free of symptoms till 1971, when the tumour recurred. He had several operations during 1971–8, and after each operation there was partial regression of proptosis. He was known to have been diabetic for the previous 20 years.

In September 1979 examination revealed the visual acuity OR 1/60. The eyeball was pushed forward and laterally. The right upper lid was completely ptotic due to repeated surgery. Ocular movements were restricted in all directions. The mass was not tender, irreducible, and nonpulsatile. The conjunctiva was chemotic and was protruding out of the palpebral fissure. The bony margins of the orbit were normal. The anterior segment was within normal limits and the fundus showed changes of diabetic retinopathy. Routine laboratory investigations were noncontributory except for a high blood sugar level. *X*-ray films of the orbit and ultrascan showed a large soft-tissue mass in the right orbit.

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Subtotal removal of the mass through an anterior orbitotomy was performed. The tumour extended deeply into the muscle cone, and its posterior limits could not be defined. One year after this operation the patient developed proptosis, severe pain, and absence of visual perception. Exenteration was carried out.

Histopathology. The specimens of tissue removed at different times showed almost similar morphological features characteristic of haemangioperi-

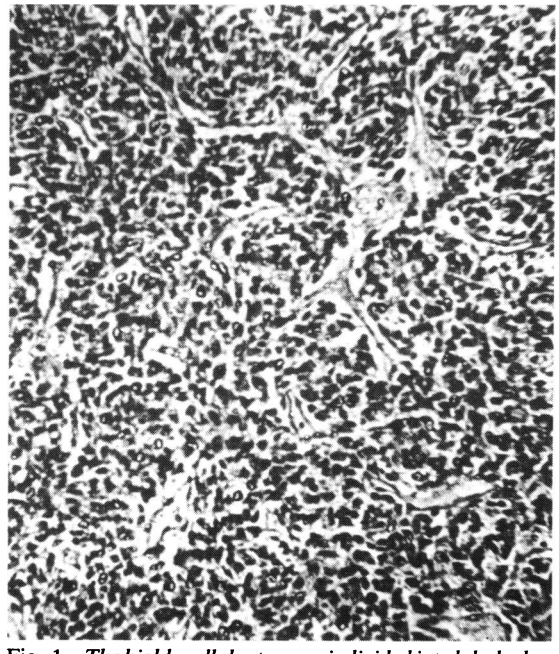

Fig. 1 *The highly cellular tumour is divided into lobules by fibrovascular septa and the cells are arranged in sheets. (H and E, ×80).*

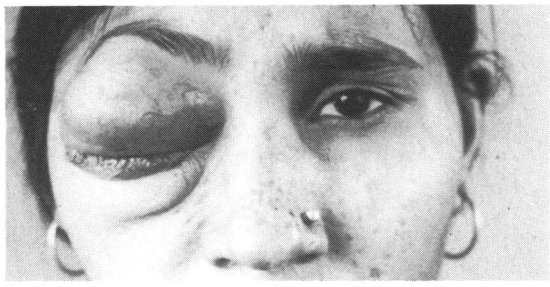

Fig. 2 *Case 3 showing proptosis.*

cytoma except for a slight increase in cellular pleomorphism in later specimens. The tumour was highly cellular and was divided into lobules by fibrovascular septa. The vessels were lined by distinct endothelium. Polyhedral cells arranged in sheets surrounded the blood vessels (Fig. 1). The cells had vesicular nuclei, pale eosinophilic cytoplasm, and indistinct cell margins. The frequency of mitotic figures varied, 5–10 per high-power field.

CASE 2

In July 1975 a 42-year-old female came with the complaint of proptosis of the left eye for one and a half years and loss of all useful vision for several weeks in the same eye, followed by proptosis. She had been operated upon elsewhere for this condition 7 months previously.

Examination revealed a blind left eye with marked proptosis, limitation of ocular movements, shallow anterior chamber, semidilated and fixed pupil, and cataractous lens. The right eye was within normal limits. Laboratory investigations and radiological study were noncontributory.

Subtotal anterior orbitotomy was performed and the mass was removed as much as possible. Postoperatively the proptosis receded, and the patient was lost to further follow-up.

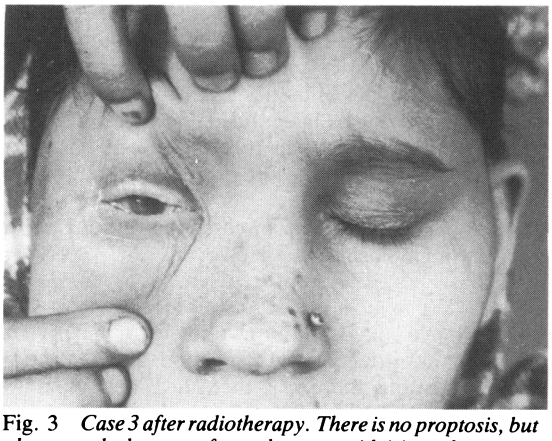

Fig. 3 *Case 3 after radiotherapy. There is no proptosis, but photograph shows perforated cornea with iris prolapse.*

Histopathology. The morphological features were similar to those of case 1.

CASE 3

A 22-year-old female was first seen in May 1979 with a history of proptosis of the right eye for one month. There was also a history of some injury to the same eye one month before the onset of symptoms.

Examination revealed an eccentric proptosis in down and out directions and fullness in the superotemporal region above the lateral canthus and below the eyebrow (Fig. 2). The orbital margins were normal. A bilobed firm and mobile mass could be felt over the superotemporal region. It was not tender and nonpulsatile, and its posterior margin could not be reached. The visual acuity was 6/18 in the affected eye. Ocular movements were restricted in all directions. There was lagophthalmos, and the conjunctiva was congested. The rest of the anterior segment structures were normal. The fundus showed temporal pallor of the disc, dilated and tortuous veins, and an oedematus macula with a dull foveal reflex. The left eye was normal.

Laboratory investigations were within normal limits. A CT scan showed a retrobulbar mass. With the above findings a clinical diagnosis of lacrimal gland tumour was made, and the patient was subjected to lateral orbitotomy. The tumour mass was removed. Postoperatively the proptosis subsided and the patient was advised periodical follow-up.

The patient returned in September 1979 for marked recurrence of proptosis with visual acuity of 3/60. She was then given radiotherapy by cobalt-60, and the proptosis totally subsided. She presented again in January 1980 with pain and redness of right eye for one month. On examination there was no proptosis and the visual acuity had fallen to only perception of light. The cornea showed a perforated ulcer with iris prolapse (Fig. 3). A diagnosis of radiation-induced perforated corneal ulcer was made, and a therapeutic penetrating keratoplasty was done. The patient was relieved of acute symptoms, but the graft became opaque in course of time. In March 1980 a firm, well circumscribed, smooth swelling, 3 cm in diameter, fixed to the pectoralis major, was detected in the right breast. The rest of the right breast, the axilla, and the supraclavicular area were normal. There was no regional lymphadenopathy. *X*-rays of chest and skeletal system were within normal limits. Excision biopsy of the breast lump was performed.

Pathology. The mass removed was 35×30 mm and was covered by a thin fibrous capsule. The cut surface had a fleshy, lobulated appearance (Fig. 4).

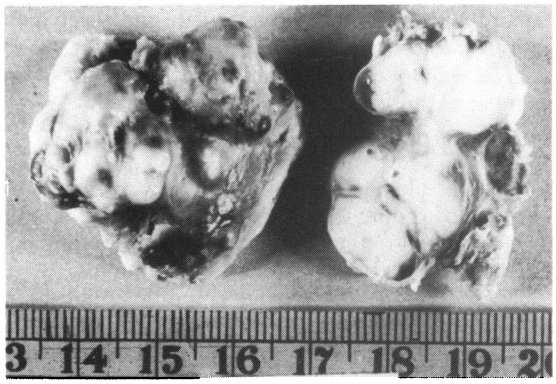

Fig. 4 *Gross specimen of case 3: left, covered by a thin capsule; right, the cut section of the specimen showing a fleshy, lobulated appearance.*

Microscopically the tumour was composed of highly cellular tissue. It was surrounded by a dense, hyalinised, fibrous tissue capsule of variable thickness on most of the surface. It was divided into lobules of varying sizes by fibrovascular septa which extended from the capsule. The cells were round to polyhedral in shape and had vesicular nuclei, with folding of the nuclear membrane and pale eosinophilic cytoplasm and indistinct cell margins. The tumour cells surrounded the vascular spaces, which had a distinct endothelial lining (Fig. 5). Reticulin stain disclosed that the cells proliferated outside the reticulin sheath of the capillaries (Fig. 6). The walls of the vascular spaces were distinct in some areas and collapsed in others. The frequency distribution of mitotic figures was very variable in different fields of the same section but on the average it was 1–5 per high-power field. The breast lump was identical to the orbital lesion (Fig. 7).

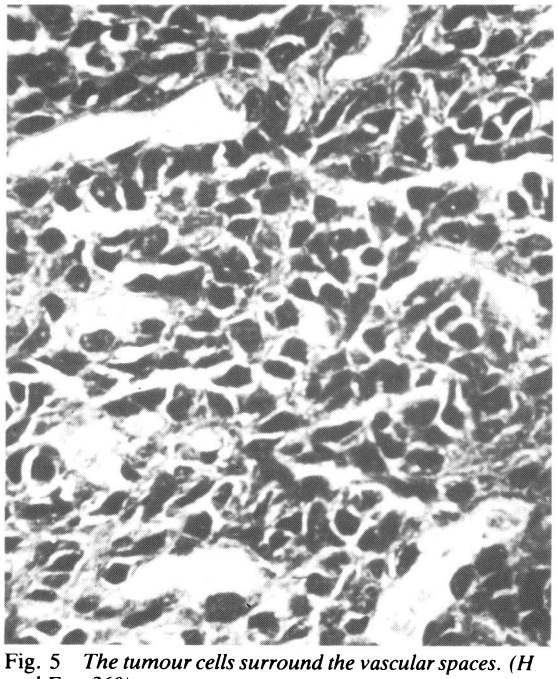

Fig. 5 *The tumour cells surround the vascular spaces. (H and E, ×360).*

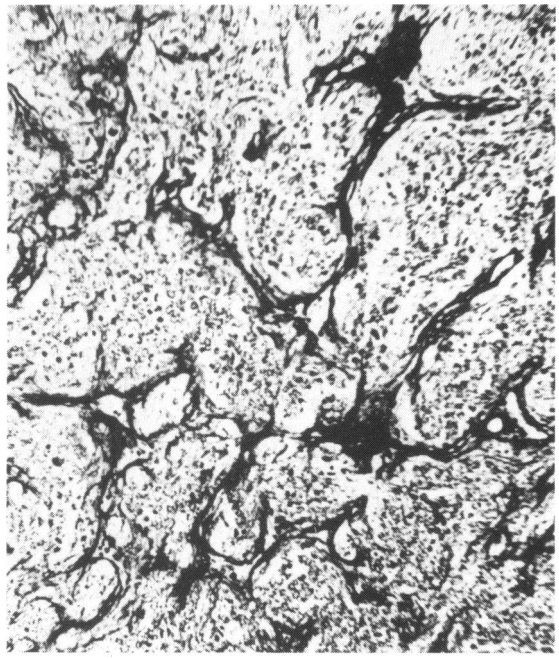

Fig. 6 *The reticulin stain emphasises the capillary element. Reticulin fibres envelop the tumour pericyte. (Reticulin, ×80).*

Discussion

Haemangiopericytoma is believed to arise from the pericytes of Zimmermann, which are found round the blood capillaries and postcapillary venules of practically all types of tissues.[7] The tumour is seldom painful. It is usually noticed because of rapid growth,[8] which was seen in all 3 of our cases, or because it appears at the site of previous trauma,[8] as in our third case.

In approximately 50% of cases the tumour either recurs or metastasises. However, the estimated metastasis rate as reported varies from 12 to 45%.[5 9–12] The most common sites for metastases are lung, bones, mediastinum, and liver.[6] Regional lymph nodes are commonly invaded. In the present series in case 3 there was metastasis to the breast within one year. This seems to be the first reported case in which the metastasis was to the breast.

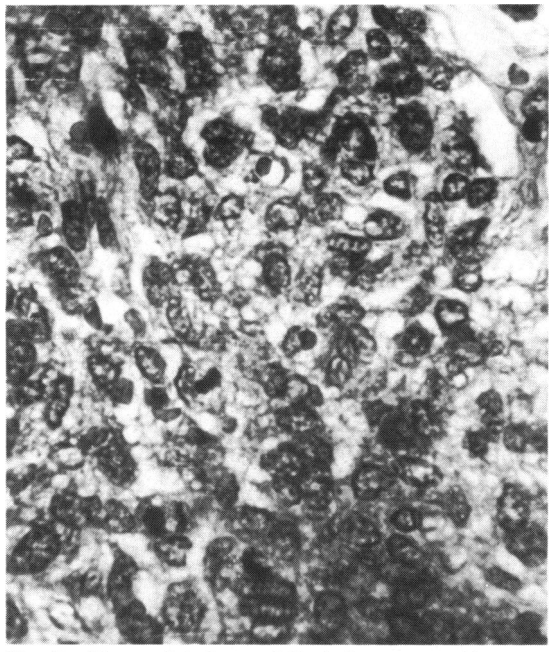

Fig. 7 *Metastasis in breast. Mitotic figures are evident. (H and E, ×320).*

The recurrence rate varies with the primary site of the tumour, being 80% for tumour within the central nervous system to 50% for the musculoskeletal system. Primary haemangiopericytoma of lung and mediastinum shows early recurrence in 36% of cases within one year, whereas that of orbit, oral, and nasal cavities and sinuses shows recurrence in 4·7% of cases within one year, 19% within 5 years, and 33% above 5 years.[1] The recurrence of primary orbital haemangiopericytoma depends also on the type of management. Backwinkel and Diddams[1] had recurrence in 22% of cases after surgery, 13·3% after radiotherapy, and 14·8% after surgery and radiation.

However, in our series the first case enjoyed a lengthy period of freedom for 22 years. The second case showed recurrence after 6 years and thereafter very frequently, while the third case had the first recurrence within 4 months.

The tumour is relatively radioresistant, and wide surgical excision is the treatment of choice.[1] As we had many problems in the first case, the third case was subjected to radiation at the first recurrence. Though the proptosis subsided, she developed radiation sequelae in the form of perforated corneal ulcer.

Orbital haemangiopericytomas are generally encapsulated, but at times show high cellularity and many mitoses that indicate the potential of the tumour for recurrence if incompletely excised.[3] Enlarged orbital dimensions in *x*-ray studies as well as encapsulation indicate a long growth period.[3] Wide surgical excision without rupture of the tumour's capsule is obligatory if recurrence is to be avoided.[3]

The prognosis for haemangiopericytoma is somewhat better than that for haemangioendothelioma, which has a metastasis rate of at least 56%.[9] The better prognosis in these cases may be due to the fact that the tumour cells proliferate outside the capillary reticulin sheath. Some workers have tried to base prognostic criteria on histopathology, depending on the degree of cellularity, cellular pleomorphism and frequency of mitotic figures, while others consider that prognostication on the basis of histopathology is misleading.[2,4] It is difficult to comment on this question on the basis of only 3 cases. However, we tend to agree with the latter workers, because the microscopic picture was quite variable from section to section and in different areas of the same section. In our cases there was hardly any morphological difference at microscopy between tissues from case 1, who had been having locally recurrent tumour for the last 30 years, and tissues from case 3, who developed recurrence and a distant metastasis within 4 months of diagnosis.

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