Intravascular papillary endothelial hyperplasia of the orbit

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SUMMARY  Intravascular papillary endothelial hyperplasia is a rare vascular benign tumour bearing some similarities to malignant angiosarcoma. The case reported here in a 20-year-old girl is of particular interest because the orbital tumour, despite its entirely benign nature, invaded the temporal fossa through the lateral bone of the orbit.

Intravascular papillary endothelial hyperplasia (IPEH) is a rare type of vascular tumour that was first described by Masson in 1923 under the name of ‘vegetant intravascular haemangioendothelioma’. Clinically it generally appears either as a subcutaneous or intramuscular nodule or in thrombosed vessels, principally haemorrhoidal veins. It has been variously described as endovascular haemangioendothelioma, intravascular angiomatosis, and intravascular papillary endothelial hyperplasia. The ocular region has rarely been affected; only 3 cases involving the eyelids and 2 the vena angularis have been reported. Three orbital tumours described under the name of angioblastoma by Martelli and considered by Wolter and Lewis to be cases of intravascular papillary endothelial hyperplasia were more probably cases of capillary haemangioma, haemangioendothelioma, or haemangiopericytoma. The present study concerns a case of IPEH which appears to be the first observed in the orbit. It is of particular interest because the tumour, despite its benign histological character, invaded the temporal fossa through the bone of the orbit.

Case history

A 20-year-old girl reported progressive exophthalmos of the right eye over a period of 1 year. The first ophthalmological examination revealed a 6 mm proptosis of the right eye which did not affect eyeball movement. Enlargement of the palpebral fissure (14 mm on the right side and 12 mm on the left) was observed. No palpable mass was noted, but the orbital rim was sensitive to touch. The fundus of the eye and the visual field were normal.

The computerised tomography (CT) scan showed a well defined, voluminous, extraconical tumour in the external anterior part of the right orbit, which had caused erosion of the external bone wall (Fig. 1). Three weeks later the patient reported headaches, malaise, dizziness, and vomiting. Although her general health was excellent, the condition of her eye had deteriorated. The proptosis had grown to 8 mm, and a slight palpebral oedema could be seen. A painful mass had developed in the region of the lacrimal gland, and a soft, nonpulsating swelling in the right temporal fossa was observed. The visual

Fig. 1  CT scan, horizontal section. The tumour is clearly visible in the right orbit between the bone, which appears eroded, and the eye and lateral rectus.
field and acuity had remained normal, but a slight papillary oedema had developed. Ocular motility was normal, and there was no evidence of abnormality of either thyroid or neurological function. Echography A showed irregular peaks in the external half of the orbit and behind the eyeball, indicating a voluminous and nonhomogeneous tumour.

A second CT scan with coronal sections taken 4 weeks after the first showed an hour-glass shaped growth in the upper external part of the orbit, with invasion of the middle and anterior parts of the temporal fossa.

A frontotemporal craniotomy of the right orbit permitted the total extirpation of an intraorbital mass and partial extirpation of an ill defined temporal tumour. Postoperative recovery was uneventful. As a result of the operation the proptosis measurement decreased (4 mm) but a slight oedema of the temporal and malar region remained. One year later the eye was found to be normal in all other respects, and the patient reported no pain or other discomfort.

**Fig. 2** Periphery of the tumour showing the capsule made of parallel lamellae of collagen representing a vascular wall, the vascular lumen, and the papillary projection into the lumen. (H and E, × 22.)

**ANATOMY AND PATHOLOGY OF TUMOUR**

The orbital tumour consisted of an oval-shaped mass of 2.0 × 1.5 × 1.5 cm, macroscopically well defined and appearing encapsulated. Microscopic examination showed that half the tumour's outer surface was covered with a smooth, regularly structured capsule, while the other half, which had been in contact with the orbital wall, was unencapsulated, irregularly structured, and contained some bone fragments. The smooth side of the tumour was constructed of parallel connective lamellae of highly variable thickness, suggesting a venous wall (Fig. 2).

The interior of the tumour was partly obliterated by a proliferation of loose connective tissue forming papillary structures (Fig. 3) that were covered by a continuous lining of flat or plump endothelial cells (Fig. 4). This vascular lumen contained a relatively unstructured mass of fibrin, old red cells, and haemosiderin pigment that was probably an old thrombus. The entire surface of the mass was covered with endothelial cells which also formed papillary projections that were frequently in contact with those extending from the outer wall of the tumour. Endothelial cells also proliferated in the connective tissue that narrowed the vascular lumen, forming a new growth of capillaries. Although frequently swollen, the endothelial cells never appeared atypical, and no mitosis was observed (Fig. 5). The stroma of the tumour was slightly coloured by periodic acid Schiff stain and alcian blue and contained few cellular elements. No cellular anomaly or mitosis was observed (Fig. 5).

Areas of dense fibrosis with abundant reticulin fibres as well as small deposits of haemosiderin were found throughout the tumour. Several foci of lymphoplasmocytic inflammatory infiltration without follicle formation were observed, particularly around newly formed capillaries. In some areas the interpapillary spaces contained a homogeneous hyaline substance that seemed to be undergoing osteoid transformation (Fig. 6).

The extraorbital tumour was much less well defined than the intraorbital part and was composed of several fragments, one of which could be seen to infiltrate the striated musculature (Fig. 7).

The most striking characteristics of the tumour were its infiltrating nature, its marked fibrogenic effect, and the abundance of lymphocytes and plasma cells surrounding its newly formed capillaries.

**Discussion**

Both the clinical evolution of the condition and the preoperative examination strongly suggested a
Fig. 3  Vascular lumen of the tumour, almost obliterated by papillary projections. (H and E, \( \times 21 \)).

Fig. 4  Endothelial lining of the papillary structure and infiltration of the loose connective tissue of the stroma by vascular spaces. (H and E, \( \times 54 \)).

Fig. 5  Detail of the endothelial lining of the papillae. All endothelial cells are flat and free of anomalies. (H and E, \( \times 358 \)).
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Intravascular papillary endothelial hyperplasia of the orbit is generally considered to be a specific form of organisation of a thrombus rather than a primary intravascular tumour complicated by a thrombus.\(^1\) In spite of the infiltrating nature of the tumour observed in the present study, the diagnosis of IPEH was confirmed by several other pathologists (Dhermy, personal communication; Font, personal communication), and a similar benign infiltrating tumour has also been described by Rosai and Ackerman.\(^1^0\) The tumour described in the present study probably developed in a dilated intraorbital vascular malformation. Its infiltration into the temporal fossa might occur by the passing of the malformation through the bone wall. Unfortunately this hypothesis could not be confirmed in the present study.

Fig. 6  Hyaline and homogeneous substance undergoing osteoid transformation in the vascular spaces between the papillary structure. (H and E, \(\times 54\)).

Fig. 7  Fibrogenic reaction in front of the tumour invading the muscle of the temporal fossa. (H and E, \(\times 54\)).
study owing to the piecemeal extirpation of the affected bone. The possibility of IPEH becoming malignant is still under discussion, and it is not inconceivable that the cases of 'malignant endovascular papillary angioendothelioma' in 6 children described by Dabska had evolved from some IPEH type of tumour. The tumour observed by Dabska might, on the other hand, represent a specific type of neoplasm which could be classified between IPEH and angiosarcomas from the point of view of malignancy.

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