Haemangiopericytoma of the lacrimal sac: a case report

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SUMMARY A primary haemangiopericytoma of the lacrimal sac in a 45-year-old male is reported. The neoplasm, highly unusual in this location, recurred twice before being correctly identified. Total excision with surrounding tissue appears to be the treatment of choice for this tumour, the behaviour of which is often unpredictable.

After the concept of haemangiopericytoma as a tumour of pericytes was introduced by Stout and Murray in 1942,¹ its occurrence has been reported in many parts of the body, most often in the head and neck.² In the ophthalmic literature we find it reported mainly in the orbit as single case reports or as clinicopathological studies of larger series.³ To our knowledge only one case has been previously observed in the lacrimal sac.⁴

Case report

The patient, a 45-year-old male, traced his clinical history back to when he was 40. Then he began complaining of a progressively increasing right eye epiphora which led after about six months to a dacryocystorhinostomy. The patient remained free of symptoms for several months, after which the epiphora recurred and a mass appeared below the right medial canthus. The mass was for some time reducible, as if due to dilatation of the lacrimal sac; later on it acquired the clinical features of a slowly growing neoplasm. Surgical intervention 24 months after dacryocystorhinostomy revealed a tumour involving the entire lacrimal sac. It was then histologically diagnosed as a 'benign cellular schwannoma'. The neoplasm recurred one year later, necessitating a second operation. A further recurrence became apparent seven months later, at which time the patient was sent to our clinic for consultation.

Physical examination revealed a firm, non-mobile subcutaneous mass bulging below the right medial

canthus (Fig. 1). The mass was painless, and the overlying skin, not involved by the tumour, was hyperthermic by 2°C when compared with the opposite site. The CT scan showed a solid tumour with well defined contours occupying the bone defect left by the previous dacryocystorhinostomy. The tumour projected into the nasolacrimal duct, which was markedly dilated (Fig. 2). At surgery the tumour appeared under the medial palpebral ligament as a well circumscribed mass, surrounded by dense connective tissue and engaged in the nasolacrimal duct. After sectioning of the ligament the tumour was removed together with the surrounding tissue and the mucosa of the nasolacrimal duct. Infiltrating areas

Fig. 1 Clinical photograph taken at the time of the second recurrence, showing the tumour mass bulging just below the right medial canthus.

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Fig. 2 Coronal (left) and axial CT scan (top right) show the tumour filling the bone defect, but no destruction of the bony margins. On lower axial scan (bottom right) the enlarged nasolacrimal canal is seen (compare with the opposite canal on the same scan).

were not found, and the tumour mass could be easily removed in toto.

On its cut surface the neoplastic tissue was firm, greyish-white, and homogeneous, with a slightly multinodular texture. Microscopically the tumour showed a prominent vascular pattern, best appreciated with reticulin stain, with tightly packed oval or spindle shaped cells growing haphazardly around thin-walled vascular channels (Figs. 3, 4). The tumour cells were uniform, with plump nuclei, ill-

defined cytoplasmic borders, and a moderate degree of anaplasia (Fig. 5). Scattered areas were highly cellular, with four to five mitotic figures in 10 high-power fields. A fibrous capsule surrounded the entire neoplasm. Neither haemorrhage nor necrosis was observed, nor was vascular or capsular invasion. The immunohistochemical stain for S-100 protein was negative throughout the tumour tissue. The factor-VII-related antigen was present only in the endothelial cells lining the vascular spaces.

Fig. 3 The tumour cells proliferate around and between thin walled vessels and vascular spaces lined by a flattened endothelium. H and E, $\times 100$.

Fig. 4 Reticulin stain outlining the vascular structures ramifying throughout the tumour tissue, and the reticulin meshwork between the proliferating cells peripheral to the vascular channels. Gomori's method, $\times 100$.

Because of the morphological and immunohistochemical findings the diagnosis of haemangiopericytoma was suggested. We subsequently had the opportunity to review the slides of the previous histological examinations: the haemangiopericytomatous pattern was already present, though

somewhat inconspicuous, since the vascular spaces were compressed and obscured by the proliferating cells. Compared with the previous slides the present morphology showed an increased cellularity of the neoplasm, with higher mitotic index and an increase in the degree of cellular anaplasia.

Fig. 5 In the most cellular areas the tumour cells have uniform larger nuclei, are in a disorderly arrangement, and compress the vascular channels. H and E, $\times 200$.

The postoperative course was uneventful and the patient was dismissed without any further therapy. Three and a half years after the last operation he was alive and free from signs of recurrence or metastasis to distant sites.

Discussion

Haemangiopericytoma is an uncommon neoplasm, and knowledge of its clinical manifestations, biological behaviour, and treatment derives largely from cases affecting soft tissues. Its histological identification and differentiation from imitators rests entirely on adequate sampling from various areas of the tumour to demonstrate the uniform cell population and the uniform histological appearance. In the present case previous consultants had interpreted the lesion as a benign cellular schwannoma. We rejected this interpretation on the grounds of both the morphological and the immunohistochemical findings in the recurring tumour. Further, the thermographic findings gave a possible clue to the nature of the tumour, indicating its high vascularity.

The morphological distinction, or grading, of haemangiopericytomas in terms of their malignant potential is difficult and somewhat controversial. Some authors maintain that accurate grading is possible and useful,^{2,5} while others have presented evidence that the assessment of the malignant potential from morphological features is not reliable.⁶ This was also the experience of Croxatto and Font³ in their series of orbital tumours, where recurrences and metastases developed in cases classified benign or borderline.

In our case not all the morphological signs of malignancy were present, while the clinical history clearly indicated an aggressive behaviour, with recurrences within a short span of time. The first clinical symptoms, as in most lacrimal sac tumours, appeared long before the tumour growth became evident, mimicking a chronic dacryocystitis or a post inflam-

matory stenosis of the lacrimal tract, thus leading to a delay in correct diagnosis and appropriate therapy. But in our case both the clinical history and the morphology of the lesion, which changed on successive histological examinations, suggest that the neoplasm may have been benign at first, becoming malignant only later in its natural history. Such a course has also been observed by Massarelli *et al.*⁷ in a haemangiopericytoma of the parotid gland. It is also reported, however, that a borderline malignant appearance may not significantly change over a span of many years.⁸

A wide total incision of the tumour, whenever possible, appears to be the treatment of choice. The efficacy of radiotherapy and chemotherapy is not established at present. If removal is incomplete, recurrences are almost the rule. The local recurrence rate for this tumour varies from 17 to 80% and is dependent also on the site of the lesion: in the orbit it approaches 30%.² However, long follow-up periods are mandatory, as the tumour may recur as long as 20 or more years after the initial surgical treatment.

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