Sir,

Intravascular papillary endothelial hyperplasia (IPEH) mimicking a lacrimal sac mass

A patient with epiphora and a medial canthus mass below the medial canthal tendon most often has a lacrimal sac mucocoele. We present the case of a woman with epiphora and a painless medial canthal swelling who had an exogenous benign vascular tumour anterior to the lacrimal sac. We discuss the differential diagnosis of medial canthal masses.

Case report

A 59-year-old woman presented with a 16-month history of right epiphora, more marked than on the left. She had a non-reducible mass below the right medial canthal tendon. This was non-tender, firm and rounded and it seemed to arise from the lacrimal fossa. The overlying soft tissue was normal, with no signs of inflammation. There was a slight bluish colour visible through the skin and orbicularis (Fig. 1a).

Lacrimal investigations included syringing, dacryocystography and nuclear lacrimal scintigraphy. The syringing on the right showed a partial block with some regurgitation but patent with pressure. The left syringing was entirely patent. The dacryocystogram and nuclear lacrimal scintigraphy showed findings consistent with right upper nasolacrimal duct stenosis, but no intra-sac lesion.

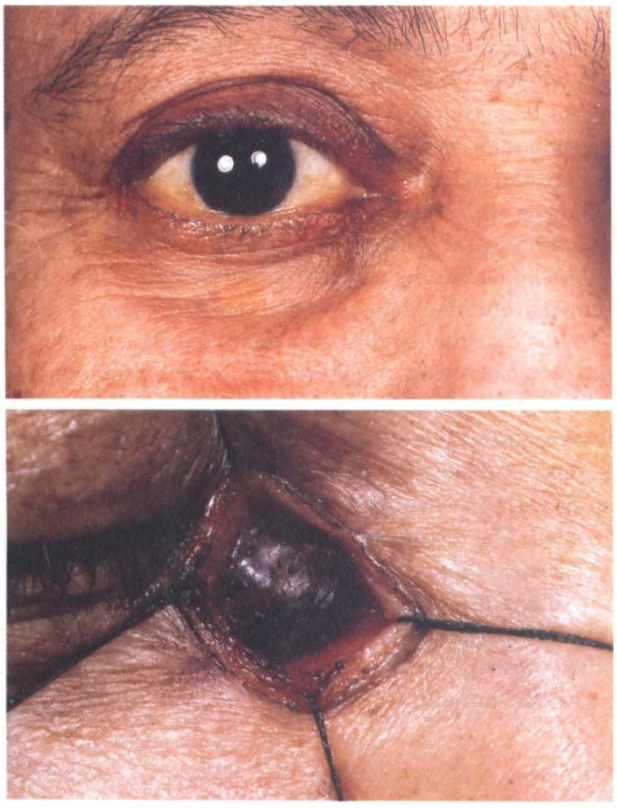

Fig. 1. *Above: A small bluish-coloured lump is seen inferior to the right medial canthal tendon. Below: Per-operatively the skin and orbicularis are reflected, revealing a vascular tumour.*

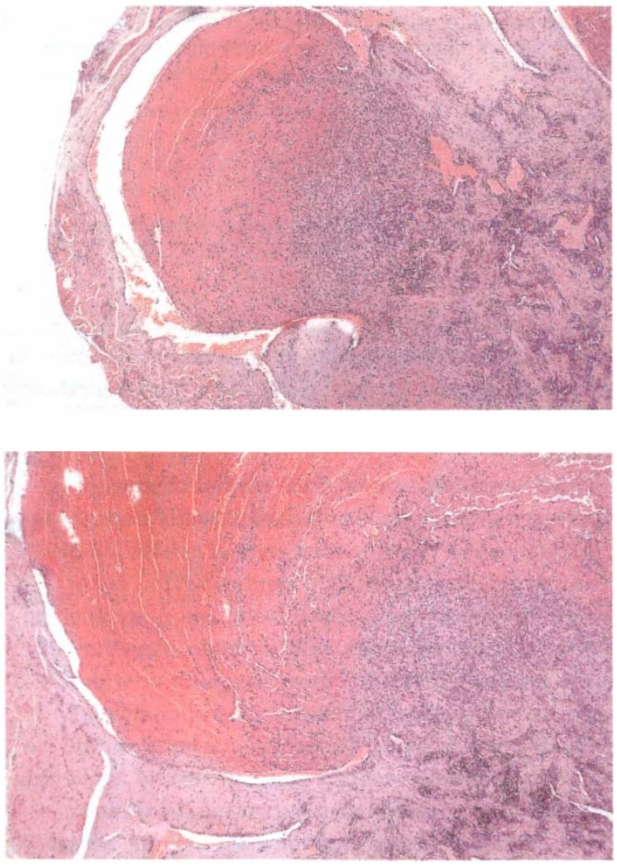

Fig. 2. *Above: Dilated vessel containing focal papillary growth composed of proliferating endothelial cells with overlying thrombus (haematoxylin and eosin, original magnification ×4). Below: Higher magnification showing intraluminal papillary growth and adjacent chronic inflammation and thrombus (haematoxylin and eosin, original magnification ×40).*

A diagnosis of functional nasolacrimal duct obstruction was made. This did not fully explain the medial canthal mass and therefore exploratory surgery was undertaken via a tear trough incision. This incision extended from just below the medial canthal tendon for approximately 15 mm inferolateral in the tear trough line, immediately overlying the mass.

A dark blue purplish vascular tumour was shelled out intact from anterior to the lacrimal sac (Fig. 1b). Macroscopically the lump was dark blue, firm and appeared to consist of medium-sized blood vessels, consistent with a haemangioma containing thrombosis. Histological examination showed intravascular papillary endothelial hyperplasia in which medium-sized vessels containing organised thrombus and adjacent intraluminal papillary projections were found. There was no infiltration into surrounding tissue (Fig. 2a, b).

The patient made an uneventful recovery. Two years later, mild functional epiphora persisted, due to functional nasolacrimal duct obstruction as indicated by the resistance on syringing and imaging.

Comment

A mass at the medial canthus below the medial canthal tendon, in the presence of epiphora, is presumed to arise from the lacrimal sac, most likely a mucocoele secondary

Table 1. *Exogenous lacrimal sac masses*

Haemangioma	Neurofibroma
Lymphoma	Fibrous dysplasia
Osteoma	Idiopathic orbital inflammation
Wegener's granulomatosis	Squamous cell carcinoma
Adenoid cystic carcinoma	Basal cell carcinoma
Sebaceous gland carcinoma	Inverted papilloma
Malignant metastases from breast, prostate or melanoma	

to nasolacrimal duct obstruction. This patient's lesion could easily have been mistaken for a non-expressible lacrimal mucocoele. However, the slight blue coloration raised our clinical suspicion that the lesion was another pathology.

When assessing a mass at the medial canthus, it is important to decide firstly whether it arises from above or below the line of the medial canthal tendon. Masses arising from above the medial canthal tendon are unlikely to be from the lacrimal fossa, and more likely to be from the ethmoid sinus, orbit or brain. An ethmoid mucocoele has a firm smooth surface with or without overlying soft tissue inflammation. A superomedial orbital dermoid is usually attached to a bony suture. An infected epidermoid cyst can cause an abscess above the medial canthal tendon. Other superomedial orbital masses presenting as an anterior mass include orbital cavernous haemangioma, lymphoma, lymphangioma, haemangiopericytoma, varix and lipoma. A midline encephalocoele appears as a soft mass arising from the medial orbital wall.

Masses below the medial canthal tendon are most commonly associated with the lacrimal sac and may or may not be inflamed. They should be divided into those that are exogenous (as in this case) or endogenous to the lacrimal sac.[1–3] Some lesions may start as exogenous then become endogenous (i.e. invade into the sac from the surrounding tissues, e.g. maxillary sinus tumour). Masses that originate below the medial canthal tendon will rarely extend above the tendon (apart from a malignant lacrimal sac tumour); instead they will expand inferolaterally in the direction of least resistance from the overlying soft tissue. The lacrimal sac lies anterior to the orbital septum, and therefore orbital invasion only occurs rarely (e.g. recurrent medial canthal basal cell carcinoma).

Exogenous masses such as squamous cell carcinoma and lymphoma may arise from the maxillary sinus, and a CT scan may be required to establish this. Exogenous masses may mechanically impede lacrimal drainage by compressing and/or invading the lacrimal sac (Table 1).

The commonest endogenous lacrimal sac mass is a mucocoele. These can be expressible (reducible), or non-expressible (non-reducible) if the valve of Rosenmuller is inflamed with the common canalicular opening into the sac compressed. Lacrimal sac tumours are rare but important as malignancies in this area can have a poor prognosis due to their aggressive behaviour. Most lacrimal sac tumours are primary and about 75% are malignant. Most tumours are epithelial in origin and include papilloma, squamous and transitional cell carcinoma and oncocytic adenoma. Mesenchymal tumours are less common and include haemangioma, haemangiopericytoma, lipoma and angiosarcoma. Other tumours include lacrimal sac lymphoma and malignant melanoma (rare).[2,3]

Intravascular papillary endothelial hyperplasia (IPEH) is a rare benign vascular tumour described by Masson in 1923 and originally called 'Masson's vegetant intravascular haemangioendothelioma'.[4] It has a typical microscopic appearance with intravascular fibrous papillary fronds, covered by pleomorphic endothelium, with a variable amount of thrombus. It has a predilection for the head and neck region[4–13] and has been described in the lids,[8,13] orbit,[7] scalp,[11] nose, paranasal sinuses,[12] oropharynx, larynx, spinal cord[10] and even the brain. Other described locations include the internal auditory canal and fallopian tube.[9] It most commonly presents as a small firm bluish lesion in the subcutaneous tissue, which grows slowly and may be tender to touch. In a number of cases, trauma has been thought to be a contributing factor and in others a pre-existing vascular lesion has been noted. Our patient had no recollection of facial trauma or pre-existing lesion. There is controversy as to whether IPEH represents a primary neoplastic process or a secondary proliferation in the organisation of a thrombus. Most authors cite a lack of recurrence as the eventual outcome and conclude that it is not a malignant or pre-malignant condition. The histopathological differential diagnosis includes angiosarcoma but the vegetant intravascular haemangioendothelioma is entirely intraluminal.

In this case the surgical approach was via a skin–orbicularis tear trough incision over the tumour, which also allows access to the lacrimal area if needed. We recommend this approach for exploration of lacrimal area masses (below the medial canthal tendon) as it will also provide excellent access for dacryocystorhinostomy if needed. The resultant scar is barely visible.

This case highlights the importance of considering the wide differential diagnosis of a mass at the medial canthus, especially when there is only partial lacrimal outflow obstruction. Fortunately our patient only had a rare benign tumour.

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