woman aged 42 years who presented with a sore eye; the other in a male whose age and presentation were not stated. Pathological examination of biopsies in both showed 'cysts' in which an initial diagnosis of 'concretions' in the former and 'grains' in the second was made. The third was a man aged 94 years who presented with a senile ectropion of the eyelid. Biopsy revealed a cyst and initially a diagnosis of nematode infestation was made.

The appearance of the structures in the current case, along with the lack of any organoid structure resembling known parasitic infestations in association with a cyst of Moll's gland, satisfies the criteria for Liesegang rings.

When considering the differential diagnosis of unusual cystic lesions of the eyelid the oculopathologist needs to be aware of Liesegang rings as a separate entity and not related to parasites.

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Surgical management of primary localised conjunctival amyloidosis causing ptosis

EDITOR,—Primary localised conjunctival amyloidosis characteristically presents as an enlargement or swelling of the eyelid which is frequently associated with spontaneous haemorrhage.¹ The condition usually presents in middle age and is often bilateral. Typically, there are solitary or multiple, firm, rubbery, well vascularised, waxy appearing, painless, fusiform or polypoid subconjunctival elevations. If the tarsal plate and adjacent muscular and subcutaneous layers are infiltrated, the eyelid may be diffusely thickened. Ptosis may result from the weight of the amyloid and infiltration or stretching of the levator palpebrae superioris complex.

The surgical management for this condition has not been well described and indeed surgery is often avoided for fear of severe haemorrhage. We present a case in which a meticulous excision of the amyloid mass using a microdiathermy needle, combined with an advancement of the levator aponeurosis, provided a good functional and cosmetic result.

CASE REPORT

A 70-year-old white woman was referred for a second opinion on her longstanding bilateral ptosis. In 1982 the patient had developed a gradual swelling of her left upper eyelid which gradually resolved over a 2 week period and

Figure 1 At presentation the left upper lid is ptotic and has a bulky appearance as a result of amyloid deposition.

was associated with the development of a bruise in the same region. Over the course of the next 3 years the patient suffered recurrent spontaneous haemorrhages into both upper lids with extensive bruising. Both lids became increasingly swollen and a secondary ptosis developed. In 1985 a left posterior approach levator resection was attempted but the procedure was abandoned because of severe haemorrhage. In 1990 a Fasanella-Servat procedure was performed by another surgeon on the left upper lid as the ptosis was obscuring the pupil. This procedure gave a functional improvement for 3 years. Histopathological examination of the excised tarsconjunctiva showed the presence of amyloid tissue.

On examination the patient had a bilateral ptosis with a palpebral aperture of only 3 mm on the left side and 5 mm on the right. She had a bulky appearance to both upper lids with firm, painless, rubbery swellings palpable in and above the upper tarsal plates. The swelling was more extensive in the left upper lid (Fig 1). There was evidence of subcutaneous haemorrhage in and around the left upper lid. The diffuse thickening of the upper lids made eyelid eversion difficult. Levator function was 8 mm on the left side and 10 mm on the right.

The patient underwent an exploration of the left upper lid via a skin crease incision under general anaesthetic. The surgery was performed using a 'Colorado' microdiathermy needle which permitted excellent dissection of the tissues with minimal bleeding. The amyloid material was exposed and dissected from the underlying tarsus and conjunctiva. The levator aponeurosis was involved in the lower two thirds. This was excised and the remaining healthy levator aponeurosis advanced onto the tarsal plate. The skin incision was closed and a pressure dressing applied for 24 hours.

Histopathological examination and immunohistochemical analysis of the material removed at surgery confirmed that the mass was composed of AL type amyloid tissue which has previously been identified in deposits of primary localised amyloidosis.^{2,3} Positive staining occurred with antibodies to kappa and lambda light chains.

Postoperatively the patient made an uneventful recovery. The left palpebral aper-

Figure 2 Twelve months postoperatively functional and cosmetic improvement are apparent.

ture measured 7 mm. There was no lagophthalmos. Both the functional and cosmetic results were good. The patient remains unchanged after 12 months' follow up (Fig 2).

COMMENT

In the ophthalmic literature there has been little discussion of the management options and operative approaches for this condition. Small, localised lesions have previously been treated by en bloc resection, but in advanced cases surgery is often not attempted because of the potential risk of haemorrhage from the fragile blood vessels typically associated with the lesions, and the extent of infiltration rendering resection impractical. Recently, Patrinely and Koch described a method of treating advanced amyloid deposits by preserving the anatomical planes of the eyelid and debulking the amyloid mass with a spooned curette.⁴ This method appears to be effective in reducing the size of the amyloid mass in advanced cases.

The above case report illustrates that resection of large areas of amyloid is technically possible with preservation of normal eyelid tissue, and avoidance of haemorrhagic sequelae. At the same time it is possible to correct a secondary ptosis safely and satisfactorily.

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Orbital varix in Grönblad-Strandberg syndrome

EDITOR,—Pseudoxanthoma elasticum (PXE) is an uncommon hereditary disorder of connective tissue, characterised by fragmentation of elastic fibres in various tissues. This abnormality accounts for its most common manifestations, including angioid streaks (breaks in Bruch's membrane), reticulated skin lesions (plucked chicken appearance in neck and axillae), and vascular abnormalities.¹ The association of PXE and angioid streaks is known as Grönblad-Strandberg syndrome.²

Orbital varices are venous malformations characterised by a weakened vessel wall and often by an interrupted elastica.³ Their combination with PXE has never been reported before. We describe a case of Grönblad-Strandberg syndrome, where an orbital varix was the presenting sign.

CASE REPORT

In October 1993 a 22-year-old woman was referred for the evaluation of a painless superior medial mass, arisen since the age of 4

Fig 1A

Fig 1B

Figure 1 Clinical photograph at presentation. A superior medial mass is apparent in the right orbit (A), increasing in the prone position (B).

years in the right orbit, with no inflammatory signs. The mass augmented when the patient bent over or coughed. On examination, a bluish soft mass was apparent in the medial aspect of the right superior eyelid, becoming more evident with Valsalva manoeuvre or prone position (Fig 1). No proptosis was observed. Best corrected visual acuity was 15/20 in the right eye and 20/20 in the left eye. Intraocular pressure was 15 mm Hg in both eyes. Fundus examination disclosed in both eyes subretinal red grey streaks,

radiating from a peripapillary ring and involving the macula in the right eye, in the classic pattern of angioid streaks. The optic discs were normal. Fluorescein and indocyanin green angiographies were negative for subretinal neovascularisation. Dermatological examination showed a reticulated skin lesion in the neck; characteristic of Grönblad-Strandberg syndrome. Electrocardiogram and blood pressure were normal.

Orbital computed tomography scan revealed a superficial rounded mass in the superior medial aspect of the right orbit, enhancing after contrast administration, and enlarging with Valsalva manoeuvre (Fig 2). Orbital venography showed direct enhancement of the lesion, confirming the diagnosis of superficial varix of the superior ophthalmic vein.

COMMENT

PXE is a systemic disorder of the elastic tissue, characterised by abnormal collagen or elastin, with calcification and fragmentation of elastic fibres in various tissues, including arteries, skin, Bruch's membrane (angioid streaks in the Grönblad-Strandberg syndrome), lamina cribrosa (optic nerve head drusen), gastrointestinal tract, and mitral valve.⁴ It is inherited in an autosomal dominant or recessive mode, and it is often diagnosed in the first decade.¹ The association of PXE with an orbital varix has not been described before, but it would not seem accidental. Indeed, orbital varices are a venous malformation caused by a constitutional mural weakness, often associated with an interrupted elastica.³

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Fig 2A

Fig 2B

Figure 2 Axial contrast computed tomography scan reveals a well delimited oval mass in the medial aspect of the right orbit, corresponding to a superficial varix of the superior ophthalmic vein (A). Mass enlargement is evident after Valsalva manoeuvre (B).

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