

Figure 1 B-scan of small linear disc drusen (arrow) in one of the subjects.

Discussion

This is the first description of development of optic drusen in related children with pseudopapilloedema.4,5 We found that reassuring symptomatology, the absence of optic nerve-sheath swelling on B-scans and a positive family history of pseudopapilloedema to be very helpful in excluding serious intracranial pathology, as well as reducing the need for lumbar punctures and CT scans.

Conflict of interest

The authors declare no conflict of interest.

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Eye (2011) 25, 1101-1102; doi:10.1038/eye.2011.95; published online 6 May 2011

Eccrine syringofibroadenoma of the eyelid in association with eye prosthesis

Eccrine syringofibroadenoma (ESFA) is a rare benign adnexal eccrine neoplasm. We report an ocular presentation with a unique location in the eyelid, associated with enucleation and long-standing prosthesis.

Case report

A 65-year-old man was referred to our eye clinic with a superomedial eyelid lesion just posterior to his lash line (Figure 1). The mass was present for 3 years and was slowly enlarging. He had undergone a left enucleation at the age of 1 year for a congenital abnormality of unknown aetiology and had a well-fitted prosthesis in situ.

The lesion was a $9 \,\mathrm{mm} \times 5 \,\mathrm{mm} \times 3 \,\mathrm{mm}$, solitary, elevated, non-tender polypoid mass on the under surface of the upper left eyelid posterior to the gray line. (Figure 1). His eye socket was unremarkable and there were no other cutaneous lesions. Microscopy of an excisional biopsy revealed downward proliferation of surface squamous epithelium, with thin strands of interconnecting epithelial cords and adjacent proliferation of conjunctival epithelium including goblet cells (Figures 2a and b). Epithelium surrounding a fibrovascular stroma was identified, and immunostaining with carcinoembryonic antigen showed



Figure 1 Solitary, elevated, non-tender polypoid ESFA on the under surface of the upper left eyelid posterior to the gray line.

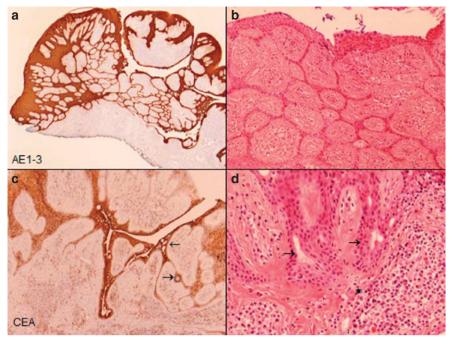


Figure 2 (a) Low-power view of polypoid lesion immunostained with epithelial marker AE1-3 to show interconnecting epithelial strands communicating with the surface. (b) Haematoxylin and eosin stained section showing higher power view of anastomosing narrow epithelial strands. (c) Low-power view immunostained with carcinoembryonic antigen to show glandular structures (arrowed). (d) High power view of dense plasma cell inflammatory infiltrate (asterisk). Duct formation (arrowed) is also visible within the proliferated epithelium. Haematoxylin and eosin.

ductal differentiation with gland lumena (Figure 2c). The underlying scarred connective tissue was densely inflamed with a mixed lymphocytic and polyclonal plasma cell inflammatory infiltrate, including Russell bodies, and a diagnosis of ESFA was made (Figure 2d). The differential pathological diagnosis for ESFA is the fibroepithelial tumour of Pinkus or pseudoepitheliomatous hyperplasia. The prominence of plasma cells has been previously noted. The rarity of this lesion, its occurrence in association with a longstanding prosthetic eye and dense chronic inflammation and fibrosis suggests that it may be reactive in nature.

Comment

First described by Mascaro in 1963, ESFAs, which can be associated with carcinomas, are rare, usually solitary, cutaneous lesions, mostly seen on the palms and soles of elderly people.1 To our knowledge, solitary eyelid ESFA on the eyelid, in association with a prosthesis, has not previously been described in the literature. Chen et al² have described familial ESFA with associated ophthalmic abnormalities in both parent and children including ectropions and entropions, absence of punctae, corneal vascularisation and scarring, and absence of eyelashes and meibomian glands. However, the ESFA plaques in those cases were on the feet, toes, scrotum, thighs, and back. Multiple ESFAs associated with ectodermal dysplasia (Schopf Schulz Passarge syndrome) can also involve the eyelid, but while the hand lesions in that syndrome are ESFAs, the eyelid lesions are apocrine hidrocystomas.3 There were no features of this syndrome in our patient and family history was negative.

Conflict of interest

The authors declare no conflict of interest.

Acknowledgements

Dr J Fitzgibbon, Mercy University Hospital Cork kindly reviewed the pathology in this case.

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Eye (2011) 25, 1102-1103; doi:10.1038/eye.2011.98; published online 13 May 2011