retraction syndrome. It lends further evidence to the theory that the two have a common teratogenic aetiology in early gestation. The fact that the entropion resolved would suggest that, in the absence of any corneal or other complications, it may be possible to closely monitor congenital entropia rather than perform immediate surgical intervention, as they may resolve spontaneously.

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Sir,

Necrosis of the lid due to *Pseudomonas aeruginosa*

Necrosis of the eyelids is rare and usually related to minor trauma such as electrical burns or spider bites, or, more dangerously, to infection.[1–7] In the absence of a history of trauma or infection a mitotic lesion such as a sebaceous cell carcinoma or a fungal lesion such as mucormycosis should be considered. Of infectious causes the commonest organisms are streptococci or staphylococci, but seven instances of *Pseudomonas* involvement have been reported, usually bilateral and associated with neutropenia.[8–14] We report such a case.

Case report

An 83-year-old woman with peripheral vascular disease and cardiac failure was admitted with pneumonia and a cellulitis affecting her lower leg, and treated with intravenous antibiotics. The cellulitis centred on a small punched-out ulcer on her left medial malleolus. Blood cultures revealed *Pseudomonas aeruginosa* and a coagulase-negative *Staphylococcus*, though neither were present in every bottle, and the question of contamination was raised. *Pseudomonas* was also found in the leg wound, however. Initial treatment with cefuroxime was therefore changed to gentamicin and piperacillin after sensitivity testing. The organism was sensitive to ciprofloxacin, pipericillin, gentamicin, ceftazidime, azatreonam, meropenem, neomycin and colistin. It was resistant to cefotaxime, trimethoprim, ampicillin/amoxycillin, cephadrine, chloramphenicol and tetracycline.

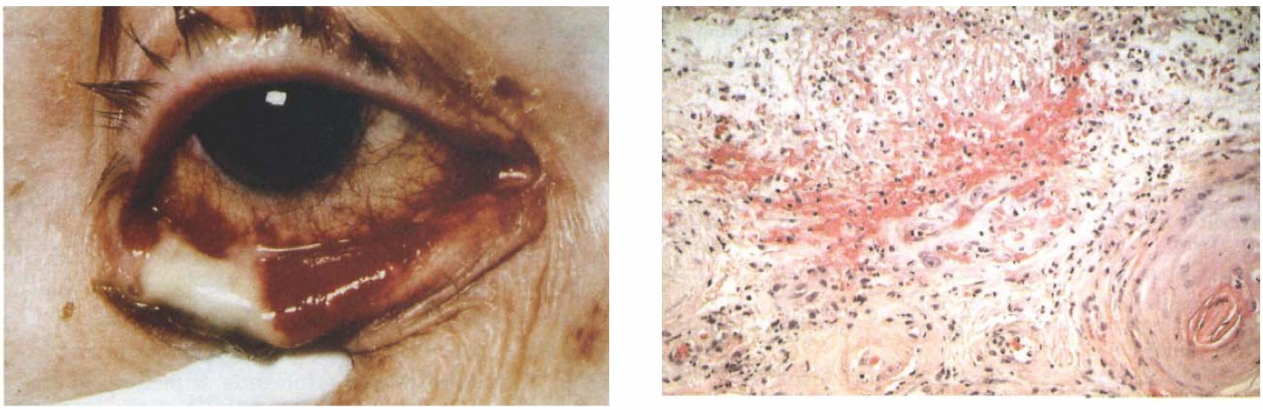
On the third day of her admission she developed a lesion on her right lower lid which was treated empirically with topical chloramphenicol and later with topical gentamicin as a swab revealed *Pseudomonas aeruginosa*, resistant to chloramphenicol. An ophthalmic review was requested on the tenth day. The patient was observed to have a grossly injected conjunctiva but, more interestingly, an area of black eschar covering the lateral third of the lid with yellowish discharge from the conjunctival surface which appeared to be frankly necrotic (Fig. 1a). Fungal involvement was felt unlikely on clinical grounds. Further swabs were taken for bacteria and fungi and topical medication changed to ciprofloxacin until a biopsy could be arranged. This was in part because it was felt the gentamicin might be exacerbating the conjunctival inflammation. The appearances began to improve. Three days later a formal full-thickness excisional wedge biopsy was performed. At biopsy the tissues looked relatively avascular. The defect was closed by primary intention with a good final result. However, the patient did not survive a developing gangrene of her leg for which she refused surgery.

The biopsy was examined at several levels (Fig. 1b). It showed an area of acute inflammation with surface ulceration and crusting and also inflammation related to occasional adnexal structures. No infecting fungus or bacteria was identified on PAS, Grocott or Gram staining and there was no evidence of tumour. Appearances were reported as non-specific but suggested the lesion may have originated within a hair follicle.

Comment

The lateral and medial palpebral arteries form two arcades in each eyelid which are thus richly supplied.[15] Healing, even after full-thickness excisions, is excellent. Infection rarely progresses because of the excellent blood supply and lymphatic drainage predominantly to the superficial parotid nodes, though debility can be permissive. Destruction of lid architecture may occur through a neoplastic process such as sebaceous cell carcinoma.

Increasingly recognised has been the occurrence of lid necrosis secondary to Group A streptococci, especially if the strain expresses pyrogenic exotoxins whilst expressing M surface protein, which facilitates invasion



(a) (b)

Fig. 1. *(a) View of the right lower lid showing purulent material on the conjunctival surface with eschar on the skin surface. (b) Micrograph demonstrating haemorrhage and deposition of fibrinoid material within the stroma of the lid and other features of acute inflammation (× 20).*

along avascular tissue planes, sometimes leading to necrotising fasciitis or necrotising myositis if involving underlying muscle.[1,2,16] This is frequently fatal even in the previously healthy, often due to a toxic shock syndrome, but wide debridement may be life-saving.[3,4] There may be bilateral involvement, with a predilection for the upper lids. The bite of the brown recluse spider (*Loxosceles reclusa*), common in the Southern United States and Israel, may leave a typical non-healing ulcer with surrounding necrosis.[5,17,18]

Pseudomonal necrosis of the lid has been reported seven times before: twice unilaterally and all with an initial neutropenia.[8–14] In one case of pseudomonal lid necrosis and one of streptococcal origin a necrotising vasculitis was thought to involve the retina or choroid also, with cotton wool spots and other signs of ischaemia being observed.[8,19] Two cases are reported as being consistent with ecthyma gangrenosum, a cutaneous lesion pathognomonic of *Pseudomonas bacteraemia*, in which a bulla develops into a necrotic ulcer covered by eschar and usually with an associated vasculitis.[9,10,20,21] Eschar was observed in our case and a bacteraemia was already suspected. She was neutropenic at the time the lid problem arose. In our case there was no definite evidence of vasculitis but there was evidence of haemorrhage and deposition of fibrinoid material in the lid stroma. Any apparently necrotic lesion of the lids in the absence of a history of trauma warrants a careful search for dangerous infective processes, many of which carry a high mortality. A full blood count and blood culture is indicated and management should be prompt. The eye itself may be involved.

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Sir,

Ptosis repair in a patient with oculopharyngeal dystrophy: brow suspension using autogenous fascia lata by spinal anaesthesia

Oculopharyngeal muscular dystrophy (OPMD) is an inherited myopathy which presents later in life with progressive ptosis and dysphagia. Beard's guidelines state that patients with less than 5 mm of levator function should undergo frontalis suspension.[1] Autogenous fascia lata is the ideal tissue to use, since it has a low failure rate and small risk of infection.[2] However, general anaesthesia in patients with muscular dystrophies has increased risks, including a malignant hyperpyrexia response and delayed respiratory insufficiency requiring post-operative pulmonary ventilation.[3] Pharyngeal involvement introduces the potentially fatal risk of pulmonary aspiration during and immediately following general anaesthesia. In this report, a patient underwent a spinal anaesthetic for harvesting of fascia lata, and then frontalis brow suspension was performed under local anaesthesia.

Case report

The patient was a 54-year-old woman, whose brother had previously undergone brow suspension ptosis surgery for the same condition. She had marked chin-up compensatory head posture with vertical palpebral apertures of 3 mm bilaterally, and a levator function of 6 mm. She had significant dysphagia, related to her dystrophy, for which she had recently undergone oesophageal dilatation, and was otherwise fit and well.

In theatre a spinal anaesthetic was administered using a standard technique: intravenous access was established and a lumbar puncture was performed, with full aseptic precautions, at the lumbar 4/5 interspace, using a 25 gauge 'pencil-point' spinal needle. Hyperbaric bupivacaine 0.5% (2.5 ml) was injected into the subarachnoid space and the patient was then turned onto her right side for 10 min to allow localisation of the neural blockade to the side of intended surgery. Following confirmation of complete sensory blockade to the level of the tenth thoracic dermatome, surgery was allowed to proceed within 15 min of injection. Fascia lata from the patient's right thigh was then stripped using a Moseley fasciotome, prepared, and a brow suspension performed using the Crawford method. Post-operatively the compensatory head posture was eliminated and the palpebral apertures were 5 mm on the right, 6 mm on the left.

Comment

OPMD is an autosomal dominantly inherited disease, caused by the variable expansion of a (GCG) repeat in the poly(A) binding protein 2 (PAB2) gene on chromosome 14q11.[4] In muscle biopsies, characteristic intranuclear filaments are found.[5] Clinically, progressive dysphagia occurs, which may require myotomy or sphincter dilatation. Proximal limb muscle weakness has also been reported.[1] Bilateral, symmetrical, progressive ptosis due to levator weakness becomes apparent in the fifth decade. Generally ophthalmoplegia is rare, and orbicularis function remains good, as does Bell's reflex. This means that the risk of corneal complications is less than for other myopathies involving the extraocular muscles. Molgat and Rodrigue[1] recommend waiting until the upper lid approaches the visual axis before operating, so that the need for further surgery as the disease progresses is delayed until late in life.

Various anaesthetic complications have been described in patients with myopathies, including cardiac arrest, rhabdomyolysis, difficulty clearing sputa and extreme sensitivity to non-depolarising muscle relaxants. In the case reported there was a significant risk of aspiration of gastric contents in the peri-operative period. Local anaesthesia is therefore recommended whenever possible. There are several reports of patients with muscular dystrophies being given spinal anaesthetics for lower limb and gynaecological surgery.[6] The injection of a small volume of local anaesthetic into the subarachnoid space in the lower lumbar region (well below the termination of the spinal cord) will produce an effective motor and sensory blockade, usually within 20 min. Use of hyperbaric preparations (with a higher specific gravity than cerebrospinal fluid) allows limitation of the level of the block to the lumbar and sacral spine unilaterally if care is taken to position appropriately, thus reducing the risk of respiratory impairment and sympathetic nervous blockade that would result from higher levels of neural blockade.

The widespread use of fine gauge 'pencil-point' needles has reduced the degree of cerebrospinal fluid leak following spinal anaesthesia. This in turn allows early mobility as soon as normal motor power in the lower limbs has returned (usually within a few hours), without a significant risk of post-dural puncture headache. Furthermore, return home on the day of surgery may be possible.[7]

Anaesthetists will often exercise caution in the use of spinal anaesthesia in patients with progressive neuromuscular diseases – a result of fear of litigation rather than any proven link between the choice of anaesthetic and the subsequent long-term progression of the disease. Although the technique may be considered safe, it is important that the anaesthetist is aware of the