Case 3

A 44-year-old man presented with a 1 week history of a painful right eye. He was also a soft contact lens wearer, removing these after 12 hours of wear and cleaning with Sauflon saline combined with Softab. His visual acuity was 6/18 and numerous subepithelial corneal opacities were noted on examination. A diagnosis of adenoviral keratitis was made and he was treated with topical chloramphenicol three times a day. One week later he presented with increased pain in the same eye accompanied by photophobia; his vision was unchanged. On examination he had a dendriform epithelial lesion and in view of the history a provisional diagnosis of *Acanthamoeba* keratitis was made and he was seen by the corneal team the same day. *Acanthamoeba* infection was confirmed by culture of a corneal scrape. He was commenced on topical neomycin and propamidine 2 hourly. He did not improve and 16 days later his vision had reduced to 6/60. Itraconazole 100 mg per day was added and later increased to 200 mg o.d., to which he showed a favourable response. Within 1 week he had a marked symptomatic improvement and at review 2 weeks later his vision was 6/12 with a quiet eye.

Discussion

Acanthamoeba infection of the eye, first reported in 1974,[3] causes a severe painful, sometimes devastating keratitis, and often eludes diagnosis. It is frequently misdiagnosed as herpes simplex infection and early signs may be non-specific.[4] *Acanthamoeba* keratitis cannot be diagnosed reliably from clinical findings in every case and successful medical treatment depends on initiating therapy early in the disease process. Bacon *et al.*[1] found that treatment within 1 month of onset results in a lower morbidity and a good visual outcome.

We suggest that *Acanthamoeba* keratitis can masquerade as adenoviral disease, as well as herpes simplex keratitis. Early adenoviral infection is characterised by diffuse punctate epithelial keratitis. Focal subepithelial opacities develop later beneath the epithelial lesions and are thought to represent immune responses to the adenovirus. Subepithelial infiltrates in *Acanthamoeba* keratitis have previously been described, but occurring much later in the disease process.[5] It is important to note that subepithelial opacification in adenoviral keratoconjunctivitis is unusual after 6–9 days. The occurrence of subepithelial opacities in the cornea of a soft contact lens wearing patient, combined with severe pain and a red eye resistant to conventional treatment, should alert the examiner to a possible diagnosis of *Acanthamoeba* keratitis. The diagnosis of adenoviral keratoconjunctivitis in a soft contact lens wearer should therefore be a diagnosis of exclusion after *Acanthamoeba* keratitis has been ruled out.

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Sir,

The Management of Keratoconus with Acute Hydrops in the Down's Syndrome and Mentally Retarded Patient

The finding of keratoconus in the Down's syndrome patient is a well-documented ocular manifestation with an incidence of up to 7%.[1,2,3] Acute hydrops in keratoconus is a rare complication but is said to occur more often in association with allergic eye disease, in Down's syndrome and in congenital rubella.[4,5] In these cases it may be triggered by repeated eye rubbing and trauma.

Acute hydrops is a common cause of blindness in the Down's syndrome patient, secondary only to cataracts and complications of cataract surgery.[2,3] In these cases further loss of vision or blindness may occur in an individual who is already mentally handicapped, adding to both the patient's problems and those of carers. Traditional treatment in these cases usually involves admission to hospital over several days, for sedation, pressure patching of the affected eye, topical treatment and, where indicated, oral acetazolamide.[6] We describe a different approach to the treatment of hydrops in the Down's syndrome and mentally retarded patient.

Case Reports

Three mentally retarded patients (patients 1 and 2 with Down's syndrome and patient 3 with severe cerebral palsy and psychomotor delay) presented to this centre in the previous 12 months with unilateral

hydrops. In one case a history of eye rubbing was elicited. Slit lamp examination was possible in only one patient. Patients 1 and 2 were first seen in the casualty department, where they were treated with topical steroids and antibiotics and the eye padded. When these cases were reviewed in the cornea clinic neither episode of acute hydrops had resolved after 3 and 4 weeks respectively. Patient 3 seen in clinic, and in acute distress, could not be examined adequately at all.

Our management was as follows. All cases were admitted to hospital as day cases and the following treatment regime carried out. All three patients had a general anaesthetic and were examined under the operating microscope. Conjunctival swabs were obtained. The integrity of the cornea was tested with fluorescein to check for ulceration and possible microbial keratitis. In patient 3 a large epithelial defect was present, and microbiological samples were taken. A subconjunctival injection of betamethasone 4 mg was made in the inferior fornix in all cases, combined with subconjunctival cefuroxime in patient 3. At the end of the microscopic examination the patients received 0.3 ml (60 units) of botulinum toxin A into the levator palpebrae muscle of the affected eye to induce a therapeutic ptosis. Discharge medication consisted only of topical steroids except in patient 3, for whom antibiotics were also prescribed.

Patients 1 and 2 were reviewed at 1 week and 2 weeks post-examination under anaesthetic (EUA) respectively. In both cases the affected eye was comfortable, a significant ptosis was observed and there was resolution of the episode of hydrops as demonstrated by the absence of conjunctival injection and resolution of stromal oedema. In patient 3 further hospital follow-up was not possible but the patient's carers confirmed that the acute pain and discomfort had resolved over a 2 week period.

In this type of management anaesthetic considerations must be taken into account. Down's syndrome patients can have cardiac problems, intravenous access can be difficult and intubation can be complex due to the shape of the face, a thick tongue and subglottic stenosis.[7,8] Therefore a competent anaesthetist aware of these problems must be present. Despite these considerations EUA is often an essential part of the management of any problem, not just ophthalmic, in the mentally retarded patient with few reported complications.

Acute hydrops develops when splits in Descemet's membrane and the endothelium of the ectatic cornea allow aqueous to enter the stroma with resulting corneal oedema.[9] In the acute phase it is always painful and associated with visual loss.[10] The oedema resolves over a period of 2–6 months and can be accompanied by scarring and flattening of the cornea.[6,9,11] A conservative approach is advocated until the oedematous process resolves,[10] but usually the central corneal scarring reduces the corrected visual acuity.[9] In the three cases described, subconjunctival injections of betamethasone and topical steroid drops were administered to reduce inflammation and stromal oedema in the acute phase. In patient 3 with a large epithelial defect, subconjunctival and topical antibotics were given to prevent microbial keratitis. Complications, although rare, can occur and include infection, perforation and neovascularisation.[9]

The use of botulinin toxin A to produce a protective ptosis is well documented,[12] and has also been used to aid in re-epithelialisation in patients undergoing epikeratoplasty.[13] It provides a practical alternative to patching in this group of patients, to relieve the pain and photophobia associated with acute hydrops.[6]

In our three patients the use of subconjunctival and topical steroids, in combination with a protective ptosis, was of benefit in effecting a speedy resolution to the episodes of hydrops, which in turn might help minimise the scarring that is a sequel to the acute phase. Therefore in this type of patient it may be beneficial in preventing further loss of vision in an individual who may already have sensory deprivation due to other ocular manifestation of Down's syndrome.[6]

Epikeratophakia for keratoconus without central scarring has been suggested in this group of patients, who can have a poorer prognosis for penetrating keratoplasty due to severe mental retardation, eye rubbing, self-traumatisation and graft rejection in the fellow eye.[14] In those with visually significant corneal scarring penetrating keratoplasty should only be considered if the patient does not exhibit eye rubbing or self-traumatisation and where there is a stable carer situation. Studies have shown that in the majority of eyes that develop severe hydrops with significant scarring penetrating keratoplasty is necessary for visual rehabilitation, but that in these eyes there is also a greater risk of rejection episodes.[9]

We feel this approach to the problem of acute hydrops in the Down's syndrome and mentally retarded patient to be desirable and practical. It ensures the patient is quickly comfortable. It also negates the need for long hospitalisation, sedation and repeated visits, as well as providing a means of accurate corneal assessment in patients who cannot co-operate with slit lamp examination.

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Sir,

The Rules of Third Nerve Palsy in Children

The rules of third nerve palsy can be useful in the diagnosis of acquired third nerve palsy. However, it is not generally recognised that there are limitations when they are applied to children. This case report illustrates this fallacy and explores the reasons for their limitations.

Case 1

A 6-year-old boy presented with a complete, left third nerve palsy associated with frontal headache, periorbital pain, cough, vomiting and pyrexia. Tenderness was elicited over the left frontal and maxillary sinuses but there were no other orbital signs or meningism. Blood films showed neutrophilia but cultures were negative. CT scan showed mucosal thickening, consistent with sinusitis, in the left ethmoidal, maxillary and sphenoidal sinuses. No intracranial lesions were seen. The patient proceeded to have magnetic resonance angiography (MRA) but no intracranial aneurysms were identified.

After a course of antibiotics, symptoms and signs of the sinusitis quickly resolved followed by a more gradual return to normal of third nerve function.

Discussion

According to the conventional rules of third nerve palsy, an aneurysm could certainly have been the cause of the child's third nerve palsy. For this reason, an MRA was felt to be necessary to exclude this possibility. Although in adults the rules of third nerve palsy can be useful as a clinical guide for indicating the likelihood of aneurysmal compression, in children they have a different significance and are of limited clinical value. The reasons for these limitations become apparent when the origins of these rules are explored.

Pain in third nerve palsy. Putative, trigeminal fibres 'hitch-hiking' in the oculomotor nerve are thought to give rise to the characteristic pain of ischaemic third nerve palsy.[1] Ischaemic pain is supposed to be dull and constant in character, in contrast to aneurysmal pain which tends to be sharp and throbbing in character.[2]

This rule distinguishes the subtle differences in the character of pain caused by ischaemic and aneurysmal lesions. Due to the subjective nature of these symptoms, this rule is of limited value even in adults.

This rule cannot be applied to children as ischaemic neuropathy is not known to occur in children.[3] Aneurysms are also rare in childhood and other more common causes of third nerve palsy such as trauma, post-viral neuropathy, raised intracranial pressure, meningitis and neoplasm can often be associated with headaches. Therefore, in a child with a painful third nerve palsy, other causes of headache should be excluded before undertaking investigations for intracranial aneurysm.

Pupil involvement in third nerve palsy. The rule states that 'when an aneurysm compresses the oculomotor nerve, the iris sphincter will be impaired'.[4]

Pupillary fibres are distributed superficially on the dorsomedial aspect of the nerve trunk and derive their blood supply mainly from pial vessels on the nerve sheath. Although they are prone to compression from aneurysms, the pupillomotor fibres are spared in ischaemic third nerve palsy when occlusion of the vasa nervorum occurs causing microinfarction of the central somatomotor fibres.[5]

This rule is also based on the contrasting clinicopathological features between ischaemia and aneurysmal compression of the third nerve and for the same reasons is therefore also not strictly applicable to children. Furthermore, pupil involvement occurs almost universally in children with third nerve palsy from non-aneurysmal causes.[3] In two large series, pupil involvement was found in 64% of adults and