

Self-inflicted corneal injuries in children with congenital corneal anaesthesia

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SUMMARY Severe corneal ulceration related to self-inflicted injury in the presence of congenital corneal anaesthesia is described in four boys under 2½ years of age. The ulcers had failed to heal until it was recognised that the children were scratching their own eyes. The application of arm splints allowed rapid healing. Although corneal ulceration is a recognised complication of congenital corneal anaesthesia, this preventable cause of the ulceration has not previously been recognised. In two cases there were isolated recurrences which healed quickly with the reapplication of splints. All four children had good vision initially, and, although there were no overt gross developmental abnormalities, two had neurological signs on detailed investigation suggesting cerebellar or brain stem malformation and one had unilateral anophthalmos, talipes equinovarus, and patent ductus arteriosus. All the children showed normal intellectual development. Whether the eye scratching behaviour was the primary cause of the ulceration or merely an aggravating factor, the identification of this abnormal behaviour is important in any child with idiopathic corneal ulceration, as even in the presence of congenital corneal anaesthesia the eyes heal quickly with effective splinting of the elbows. It is therefore important to test sensation of the cornea and face and to consider the possibility of self-inflicted injury in children with refractory corneal ulceration, as in our cases there were no other consistent diagnostic features.

Self-inflicted eye injury is an uncommon ophthalmic problem. It is recognised in blind children who rub their eyes to obtain vicarious light sensations from mechanical phosphenes and it occurs both in servicemen wishing to avoid military duty¹ and in civilian patients with psychological problems.² More severe ocular mutilation has been reported in psychotic adults³ and mentally retarded older children.⁴ Corneal ulceration is a recognised complication of congenital corneal anaesthesia,⁵⁻⁸ but self-inflicted injury has not been recognised as its cause. This paper describes four young children with congenital corneal anaesthesia and corneal ulceration which failed to heal until measures were taken to prevent them putting their fingers into their eyes.

Patients and methods

Four cases presented in a six-year period and all were

examined by a paediatric neurologist. This detailed neurological and developmental assessment revealed more associated abnormalities than were suspected at the initial clinical presentation, but all the children showed normal intellectual development.

CASE 1

A 1-year-old boy from a deprived social background was admitted to hospital with a central corneal ulcer in his right eye. He had been born with left anophthalmos associated with left sided facial microsomia, but the right eye was normal at birth. Closer examination revealed talipes equinovarus and patent ductus arteriosus. Sensation on both sides of his face appeared normal, but there was anaesthesia of the right cornea. His corneal ulcer healed slowly with topical therapy but relapsed six months later.

On his second admission he had a central corneal ulcer with hypopyon and Koch-Weeks bacillus sensitive to chloramphenicol was identified on bacteriological investigation. Despite specific antibiotic therapy the ulcer failed to heal until it was noted that

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he was rubbing the surface of the cornea with the tips of his fingers when left unattended. He was therefore placed in arm splints with the elbows extended, and this allowed the ulcer to heal rapidly, leaving a dense corneal scar which seven years later restricted vision to hand movements. He was intellectually above average.

CASE 2

A 2½-year-old boy was noted to be sticking his fingers into his eyes while being examined for suspected strabismus in the orthoptic department. The examination revealed no abnormality of vision or eye movement. Two weeks later he presented with a corneal ulcer in the right eye and associated abrasions of the forehead and periorbital skin. *Haemophilus influenzae* sensitive to chloramphenicol was isolated from the conjunctival sac, and the child was admitted to hospital for treatment, including splinting of his arms to prevent him from putting his fingers into his eyes. Detailed examination revealed traumatic depapillation of the anterior 2/3 of the tongue, with other oral signs of self-inflicted trauma. There was also partial anal atresia, cerebellar ataxia, cogwheel ocular pursuit movements, rotatory nystagmus, defective optokinetic nystagmus to both sides, bilateral corneal areflexia, and bilateral facial analgesia. Another notable feature was his fuzzy occipital hair. IQ tests were normal for his age.

This syndrome of cerebellar pathology, bilateral trigeminal anaesthesia, and peculiar occipital hair resembles the recently described cerebello-trigeminal and focal dermal dysplasia syndrome.⁹



Fig. 1 Case 2. Healing lacerations on the forehead and face after recognition and prevention of self-inflicted injury in bilateral trigeminal anaesthesia.

The corneal and skin lesions healed quickly (Fig. 1) with one brief relapse affecting the left cornea a few weeks later. A minor opacity remained in this cornea, but after four years his visual acuity remained 6/6 in each eye unaided.

CASE 3

A 16-month-old boy presented with a corneal ulcer of the right eye. He had primary hydrocephalus, and a V-P shunt had been inserted when he was 5 months old. The ulcer failed to respond to topical antibiotics, and after a few days he was admitted to hospital for more intensive treatment. Corneal sensation was absent both on the ulcerated right eye and on the apparently normal left eye. Bacterial, viral, and fungal tests and conjunctival scrapings revealed no pathogens. Serum vitamin A, serum carotene, and leucocyte ascorbic acid estimations were normal. Despite hospital treatment the corneal ulcer increased in size.

Three weeks after admission an ulcer developed on the left cornea, and self-inflicted corneal abrasions were suspected, as he was seen to be putting his fingers into both eyes and scratching his eyelids (Fig. 2). He also tended to bang his head against the side of his cot when upset. The corneal ulcers and eyelid abrasions healed rapidly after elbow splints were applied to fix his arms in extension. After three weeks at home his mother discontinued the splints as he no longer seemed interested in rubbing his eyes. When examined six weeks later the eyelid skin and corneal epithelium remained soundly healed, but there was dense stromal opacity of the right cornea with minor localised opacity of the left.

Neurological assessment confirmed bilateral trigeminal analgesia as well as jerky ocular pursuit movement and ataxia suggesting brain stem and cerebellar malformation. However, a CT scan was normal, as was serum uric acid estimation.

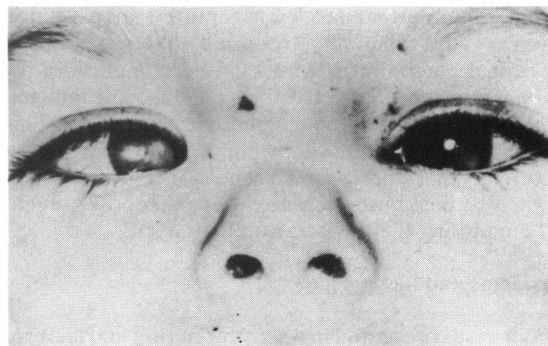


Fig. 2 Case 3. Periocular skin abrasions and bilateral corneal ulceration (stained with fluorescein). These lesions were seen to be caused by self-inflicted scratching.

Three years later there was a right convergent squint of 25° with a dense central corneal scar. The left eye recently developed a corneal ulcer precipitated once again by self-inflicted abrasion. Reapplication of arm splints allowed healing but there was residual corneal opacity. His V-P shunt was still working well, his mental development was normal for his age, and he no longer displayed head banging behaviour.

CASE 4

An 18-month-old boy was admitted to hospital because of a central corneal ulcer of the right eye which had persisted for 11 weeks despite treatment. In hospital it was noted that he poked his fingers into his right eye even when it was covered by a pad and bandage, and his mother agreed that he sometimes banged his forehead and stuck his fingers into his eyes to seek attention. Corneal sensation was deficient in both eyes, and there was anaesthesia of the forehead and scalp back to the vertex. There were no other neurological signs and a CT scan, x-ray of skull, auditory evoked responses, and blood biochemistry including vitamin A and vitamin C levels were all normal. No pathogens were isolated from the ulcer on bacteriological, viral, and fungal culture.

Arm splints were applied, but, as he could sometimes remove them or rub his eyes on other objects, goggles were strapped to his face to provide further protection (Fig. 3). It was considered that the element of attention seeking behaviour in this child made it more difficult to control his eye rubbing. With these combined measures the ulcer healed within a few days. This child appeared to be hyperactive but was intellectually normal.

Discussion

It is difficult to be certain whether the eye scratching



Fig. 3 Case 4. Goggles strapped to the face were required in addition to arm splints as he tried to rub his eyes on extraneous objects when his arms were splinted.

behaviour of these children was the primary cause of their corneal damage or whether it arose secondarily to another corneal disturbance. Case 2 was certainly seen to be putting his fingers in his eyes before the development of corneal ulceration. Moreover, these four boys with corneal anaesthesia would seem unlikely to have suffered much discomfort should there have been another primary cause of corneal ulceration and might therefore seem to have little cause to rub their eye. On the other hand that same anaesthesia may have allowed them to rub the cornea without pain when other symptoms such as watering or reduced vision had made them aware of ocular disturbance. P Wright (personal communication) has observed adult patients with acquired trigeminal anaesthesia produce excoriation of the cheek because of an insuperable feeling of needing to rub the analgesic area. Some of us may have noted a similar sensation following regional anaesthesia for dental surgery.

The cause of isolated congenital corneal anaesthesia remains obscure, though it may be associated with more widespread abnormalities such as the obvious maldevelopment of the face and skull in the Goldenhar-Gorlin syndrome,¹⁰ the generalised indifference to pain of familial dysautonomia (Riley-Day syndrome),¹¹ or as part of the condition of congenital universal insensitivity to pain.¹² None of our patients had these conditions, but it is interesting to note that two of them (cases 2 and 3) had clinical evidence of mild brain stem or cerebellar maldevelopment, features which have not previously been associated with congenital corneal anaesthesia. In case 1 it is possible that the emotional disturbance arising from a difficult maternal relationship (the mother later abandoned the child) induced either the eye rubbing behaviour or a type of temporary functional corneal anaesthesia or both. It seems that functional corneal anaesthesia can occur in adults with keratitis artefacta.² Severe and generalised self-inflicted mutilation does occur in children, but these patients usually have an underlying biochemical disorder as in the Lesch-Nyhan syndrome,¹³ the Cornelia de Lange syndrome,¹⁴ and the Gilles de la Tourette syndrome,¹⁵ but congenital corneal anaesthesia is not a feature of these cases.

Whatever the theoretical considerations, it appears that self-inflicted corneal abrasion in infants may be more common than is recognised and should always be considered as a cause of intractable corneal ulceration in this age group, especially as the presenting feature of corneal anaesthesia. There were no other consistent diagnostic features in our cases, though scratches on the face were a useful clue in two of them.

We have found that effective splinting of the child's

arms with elbows extended (permitting short, supervised periods of free mobility), allows rapid healing when combined with appropriate topical antibiotic therapy. In case 4 the attention seeking behaviour combined with corneal anaesthesia made effective protection of his corneas more difficult, and he required the addition of goggles to prevent him from rubbing his eyes on extraneous objects when his arms were splinted. Although the ulcers in all our patients healed soundly within a few days of effective protection being applied, the later relapses in cases 2 and 3 indicate the need to advise the parents of the risk of recurrences so that effective measures can be taken to limit the corneal damage if the eye scratching behaviour reappears.

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