CONGENITAL TRIGEMINAL ANAESTHESIA*

BY

G. EVERARD HEWSON†

Galway

ACQUIRED neuroparalytic keratitis following trigeminal lesions is well known. It may be due to such well-recognized causes as surgical section of the fifth nerve, for tic douloureux, syringomyelia, cranial injuries, or tumours, and is usually seen in adults. However, neurotrophic ulceration of the cornea can also occur in infants and children, in whom the trigeminal defect may be congenital, being due to a developmental anomaly in the brain stem. As few as fourteen such cases have been reported, equally distributed between the sexes.

Case Report

A female infant developed injection of the right eye with corneal infiltration at the age of one year. This improved only very slowly with local medication and similar changes appeared in the left eye 5 months later. Despite therapy with vitamin A, hypopyon ulcer followed in the right eye and uncomplicated ulceration in the left. Cauterization with carbolic acid was accompanied by bilateral tarsorraphy, but re-opening of the lids after some weeks was followed by the reappearance of corneal breakdown. Thus, tarsorraphy was repeated on both sides. Paediatric investigations failed to reveal a systemic abnormality and no signs were found of familial autonomic dysfunction, congenital anhidrotic ectodermal dysplasia, or vitamin A deficiency. The family history was unhelpful.

Eventually, activity ceased at the age of 25 months, but marked central scarring of both corneae remained. The corneae were found to be anaesthetic when the lids were opened 6 months later in April, 1959. Examination of the central nervous system did not reveal any abnormality apart from bilateral loss of pain sensation on the forehead with absence of corneal reflexes. The diagnosis of congenital trigeminal anaesthesia was suggested by Dr. S. P. Meadows.

Diminished reaction to pain is described in cases of familial autonomic dysfunction, but this condition was excluded once more despite the apparently decreased tear formation. Schirmer’s Test 1 gave wetting of filter paper to 1 mm. in the right eye and 5 mm. in the left, while physical stimulation inside the anterior nares failed to augment secretion. However, notwithstanding this apparently poor flow of tears from the lacrimal glands, there was no staining with rose bengal, which confirmed the mother’s report of normal lacrimation during crying. This suggests a reflex hypossecretion (Lyle, 1958). Bernd (1930) stressed the presence of tear formation in a patient who wet Schirmer papers to 30 mm. within 90 seconds of irritating the nasal mucosa.

No further ulceration has occurred and the eyes now appear to be normal apart from the corneal scars (Figs 1 and 2, opposite), which cause opacification in the anterior two-thirds of the thickened right cornea and in the anterior half of the attenuated left cornea. The irides are hazel in colour (Sutton, 1959) and there is no hypertrophy of the eyelashes (Duke-Elder, 1952). The present treatment consists of instillation of methyl cellulose drops (Lyle, 1958).

* Received for publication July 16, 1962.
† Late Chief Clinical Assistant, Moorfields Eye Hospital, London, E.C.1.
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Fig. 1.—Right. Fig. 2.—Left.

Discussion

Ulceration of the cornea is always a serious finding, the more so in infants. A recent report on the fatal dissemination of Pseudomonas infection from the eyes of premature babies (Burns and Rhodes, 1961) brings to mind the importance of ensuring the sterility of fluorescein drops when staining corneal ulcers in infants.

The usual causes are herpes simplex infection and simple trauma. The first condition is easily recognized and traumatic abrasions usually heal within weeks, despite the inevitable lack of co-operation from these young patients. However, notwithstanding patent nasolacrimal ducts and negative laboratory reports, keratitis with or without staining may persist and so raise suspicion of a systemic disorder. In this event, more careful supervision is required, especially of the unaffected eye.

Serious deficiency of vitamin A is exceptional in the British Isles, where systemic disorders associated with corneal ulceration include familial autonomic dysfunction, congenital trigeminal anaesthesia, and, possibly congenital anhidrotic ectodermal dysplasia (Vail, 1957; Segall, 1955; Rambo, 1958; Berenberg, 1958; Liebman, 1958). Familial autonomic dysfunction causes corneal ulceration in one-third of affected children, who may be Jewish, intelligent or retarded, with decreased muscle tone, shuffling gait, excessive sweating, drooling, and emotional upsets with erythematosus blotching of the skin and paroxysmal hypertension. These cases are usually recognized at about the age of 2 years because of difficulty in swallowing, cyclical vomiting, and absence of tear secretion. Lacrimation cannot be induced by external stimulation and there is a general indifference to pain with hypoaesthesia of the corneae. Kayser (1921) described bilateral congenital neuroparalytic keratitis which commenced at 19 days after birth in a boy who had difficulty in swallowing and chewing, salivated excessively, was retarded in development, could not stand or walk, and eventually died of inhalation pneumonia at the age of 3½ years after repeated attacks of asthma. In addition, the absence of reflex and psychic tear secretion suggests the possibility of familial autonomic dysfunction.
Congenital anhidrotic ectodermal dysplasia may be sex-linked or dominant. The fully-developed picture shows defective teeth, fine scanty hair, lack of sweating, saddle nose with chronic rhinitis, cataracts, and defective tear secretion. However, corneal ulceration does not arise solely from lack of tears.

Congenital neuroparalytic keratitis has also been described by Lawford (1907) one case, Schenk (1958) two cases, and Pillat (1949) one case. These patients were all over 5 years old. Lawford’s case may represent a near-minimal defect because lack of sensation was limited to the conjunctival sacs. Although he described ulceration of one eye only, the other cornea may subsequently have broken down, as the sensory loss was bilateral. In one of Schenk’s cases, anaesthesia and ulceration were limited to the right side. On the other hand, in Pillat’s case, there was symmetrical hypoplasia of the hair and the front portion of the temporal muscles in addition to the corneal lesions. The condition may be part of more widespread disorder, perhaps as a manifestation of status dysrhapicus (Schenk, 1958; Verrey and Jéquier, 1949). The last-named authors described the onset, between 18 months and 2 years of age, in four cases of familial neuroparalytic keratitis, with anaesthesia and hypoaesthesia in the area of the ophthalmic division of the trigeminal nerve. The family tree showed the inheritance of multiple defects, e.g. neurological lesions, ptosis, and colour blindness. Two boys aged 5 and 6 months were reported by Segall (1955). Both suffered from vertebral and other congenital defects, but malnutrition may have been an aggravating factor in causing corneal breakdown.

Apart from the loss of sensation in the forehead and conjunctival sacs, Bernd (1930) found no other abnormality in a patient whose left eye was affected at 9 months and the right at 8 years. He refers to a similar case described by van Millingen and another by Hirschberg.

The importance of recognizing the systemic implications of corneal ulceration in infants lies in expediting tarsorrhaphy, not only to the affected eye but also to its fellow at the slightest sign of corneal infiltration. Indeed, prophylactic tarsorrhaphy should be applied to the unaffected eye as soon as one is convinced of the possibility of bilateral involvement. Of fourteen cases described in the literature, only one showed unilateral sensory loss with ulceration of the ipsilateral cornea (Schenk, 1958*); in just one other with bilateral hypoaesthesia the corneal ulceration was reported on one side only (Lawford, 1907).

Perforation or phthisis bulbi may be the end-result, although not all cases are so severe. Furthermore, reflex tearing and blepharospasm are absent and fail to signal the presence of corneal breakdown in a “white” eye. Paramedian or lateral fusion of the lid margins should avoid amblyopia.

In this regard, the question of advising prophylactic cervical sympathec-

* Schenk actually recorded slight depression of sensibility on the left side in this case, but ignored the finding in his summary of it.
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Tomaty arises. There is good experimental evidence for the prevention of acquired neuroparalytic keratitis in cats by sympathectomy, as shown by Baker and Gottlieb (1959). They based their work on the clinical results of Dott and Harris (1952), who stated:

"Since experience has taught us to place complete reliance on cervical sympathectomy in the prevention of neurotrophic keratitis, the sparing of the ophthalmic sensory supply has receded in importance. In fact we can sensorily denervate the eye with impunity".

This does not seem to be widely accepted by ophthalmologists, although it had been practised for over 60 years (Vail, 1955). Such reluctance may be due to difficulty in accepting the idea that autonomic nerves play an important part in corneal physiology. This viewpoint is strengthened by the occasional report of neuroparalytic corneal breakdown in patients in whom the sympathetic is apparently out of action, as in a case of Wallenberg's syndrome quoted by Duke-Elder (1938). Nevertheless, the procedure would appear to deserve more serious consideration in both treatment and prophylaxis of corneal ulceration caused by trigeminal anaesthesia, whether it be congenital or acquired.

Summary

The development of neuroparalytic keratitis due to congenital trigeminal anaesthesia is described in a child aged one year. Differential diagnosis and prophylaxis are discussed.

I am indebted to Mr. Charles Cook, Moorfields Eye Hospital, who very kindly allowed me to describe his patient; to Sir W. Sheldon, National Children's Hospital, who carried out paediatric investigation; to Dr. Peter Hansell, Institute of Ophthamology, for the corneal photographs; to Mr. Phillip Harris, Royal Infirmary, Edinburgh, for permission to quote from his joint paper with Professor Norman Dott; to Mr. R. S. E. Brewerton of Shroffell's Hospital, Watford, who supplied details of early treatment and to Prof. Arnold Sorsby, Royal College of Surgeons of England, for references. I am particularly grateful to Mr. F. M. Sutherland, Librarian, British Medical Association, for his patience and assistance.

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G. Everard Hewson

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doi: 10.1136/bjo.47.5.308

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