ALACRIMIA CONGENITA*

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ALACRIMIA CONGENITA seems to be a very rare disease. We have succeeding in finding only nine cases in the literature. There is scant reference to the aetiology of the condition and the incidence of kerato-conjunctivitis sicca as a sequel is described only in single cases.

The first case of congenital absence of lacrimal secretion was described by Morton (1884):

Bertie L., *aet.* 6, has been noticed by his parents never to shed tears from the right eye. On several occasions I have seen him crying, and though the tears flowed copiously from the left eye, they never came from the right. That they were not secreted was demonstrated by drawing away the lower lid from the globe, when there was no accumulation of tears in the cul-de-sac thus formed. In the left eye the vision is normal, but in the right, even with the necessary correction of + 0.5 D sph. + 2 D cyl., it was not more than 20/50. The pupils act well and equally, and there is no impairment of colour vision or of the senses of touch, taste or smell on the right side. The boy's face is somewhat flatter and the external orbital angle and malar bone less developed on the right side than on the left. The right eye also appears smaller than the other. The right ear is slightly "lopped", and the upper part of the cartilage is thinner than on the left side. The mother has "harelip" on the right side, but there is no other history of deformity, and there are two younger children quite healthy. The lacrimal gland cannot be felt on either side, but it seems a reasonable assumption that it is congenitally absent on the right side.

Heubner (1900) described a boy, aged 1½, with healthy parents, who suffered from double-sided paralysis of the abducent, facial, and hypoglossal nerve, the left side being more affected than the right, with a complete lack of tears. At the examination post-mortem Heubner found that the cause was a congenital aplasia of the brain where the nuclei of these nerves lie. Thus the left hypoglossus, left facialis, and both abducens nuclei were lacking, and in the right hypoglossus and facialis nuclei the cells were fewer than usual.

Sommer (1903) described a child aged 2½ who had never shed tears. He came from a healthy family, and was physically and psychically normal. On examination of the eyes the conjunctiva was found to be normal. Irritation of the conjunctiva, cornea, or the nasal mucous membrane produced no secretion of tears, and the eyes became only a little moistened. The function of the salivary glands was normal.

According to Sommer there is reason to suppose that the case

* Received for publication June 14, 1950.

represents an abnormally long persistence of a physiological condition found in new-born children during the first weeks.

Coppez (1920) described a 2-year-old child with epicanthus and double-sided ptosis who had never shed tears.

Kayser (1921) reported a case of congenital trigeminal paralysis with total lack of tears. The child belonged to a family in which there appeared a series of congenital dystrophies. In this patient the eye symptoms began nine days after birth with small, round defects in the cornea healing quickly and leaving small scars. The process lasted till death at the age of $3\frac{1}{2}$ years. Conjunctiva and cornea were completely insensitive and the eyes dry. Tears had never been observed.

Ishikawa (1928) mentions a 4-year-old boy with no family history who lacked tears since birth. The conjunctiva was dry and slightly hyperaemic. The cornea was normal. Strong emotions and local irritation (*e.g.*, foreign bodies in the conjunctiva) failed to provoke tears. The case was explained as a congenital hypoplasia of the lacrimal gland.

Duke-Elder (1930) described a $5\frac{1}{2}$ -year-old child who had lacked tears since birth. During the first years there had been a viscous secretion from the eyes and photophobia. Both eyes seemed normal, apart from the cornea. In the deeper epithelial layers, and at some points penetrating into the superficial part of substantia propria, there were several small opacities, especially in the lower segments of the cornea. The corneal sensibility was normal. The lacrimal secretion, measured with Schirmer's test, gave sub-normal values. Stimulation by substances with strong odour also gave sub-normal values.

Bruce Hamilton (1940) described an 11-months-old child who completely lacked tears when crying. No moisture was present in the conjunctival sacs. The corneal sensitivity was poor and central keratitis was present in both eyes, with considerable areas of staining. The changes in the eyes quickly healed after obliteration of the lacrimal canaliculi.

Riddell (1940) described a 5-year-old girl who had never been able to cry, but here there were no other changes in the eyes.

Case Report

The present authors have recently examined a 5-year-old girl who had always lacked tears and showed symptoms of kerato-conjunctivitis sicca.

The patient's parents were healthy and not related. There were no known diseases in the family, particularly of the eyes, and the patient, an only child, born at term, had always been healthy. The mother told us that the girl had never produced tears on crying. The eyes became a little moistened, and the skin above them and on the forehead a little red. The child had never complained of eye symptoms and had no photophobia.

Ophthalmological Examination.—Fundi normal. Visual acuity 5/5. Corneal sensitivity normal. Corneae covered with a viscous secretion containing epithelial

cells. In the conjunctival sacs was found a scanty, tenacious secretion that could be drawn out in long threads. With rose bengal the conjunctiva and cornea became faintly but distinctly stained. Groups of epithelial cells with strongly coloured nuclei were also seen.

Schirmer's Test I: right, 1.5 mm.; left, 0.1 mm.

II: (NH₃) right, 3.0 mm.; left, 0 mm.

Tear-Gas Test: The test with tear-gas was made in a closed room, with four persons present besides the patient. All had an abundant flow of tears, but the patient's eyes remained dry in spite of psychical crying owing to pain in the eyes. The pain made it impossible to carry out Schirmer's test at this experiment.

The palpebral lacrimal gland was of normal size and appearance.

General and Neurological Examination

Blood: Hb. 80 per cent. R.B.C. 4.44, W.B.C. 4600. Blood picture normal. Erythrocyte sedimentation rate, 7 mm. per hour. Wassermann reaction negative.

Glucose-tolerance test, water-balance test, and basal metabolism normal.

Test Meal: No free HCl; fraction test showed free HCl.

X-Ray Examination: Cranium normal.

Oto-rhino-laryngologic Examination: Nothing abnormal. Salivary secretion normal.

Discussion

Thus in a child healthy in all respects, we found an absence of lacrimal secretion, even after irritation with tear-gas. Clinically there was the picture of kerato-conjunctivitis sicca. Microscopic examination of a piece of the conjunctiva bulbi showed oedema with early hydropic degeneration of the epithelium (Figure).

Microscopic examination of a piece of the palpebral lacrimal gland showed a normal picture. After obliteration of the lacrimal puncta the clinical signs disappeared.

Of the ten cases reported above, four have shown changes of the kerato-conjunctivitis sicca type. However it should be observed that in the first six cases the description was incomplete and belongs to a time when this disease was unknown. In four cases information is lacking, and in two it is reported that the eyes were normal. Thus it may be said that the characteristic hydropic degeneration of the conjunctival epithelium may appear even among children where the lacrimal secretion is lacking from birth. However, we do not know how long it takes before kerato-conjunctivitis sicca develops.

FIGURE.—Section of conjunctiva bulbi. Oedema with beginning of hydropic degeneration of the epithelium.

With regard to the cause of the lack of tears there are different possibilities. Morton (1884), whose case was unilateral and showed a series of facial anomalies on the affected side, considers it probable that the lacrimal gland was missing on that side. Lutz (1931) pointed out that here the cause might be an aplasia of the petrosal bone with co-existing aplasia of the nervus petrosus superficialis major. Coppez (1920) and Ishikawa (1928) believed that the condition was due to a congenital hypoplasia of the lacrimal glands. However, it has not been possible to prove this. Kayser (1921) sees the cause in a congenital paralysis of trigeminus. Heubner (1900), whose case showed paralysis of several cranial nerves, proved microscopically an aplasia of the nervous nuclei.

In our case no nervous symptoms could be proved and the lacrimal gland was microscopically normal. It is difficult to decide the cause of the lack of lacrimal secretion in this case. Apart from the missing lacrimal secretion, no hypofunction of the secretory organs, and no other morbidity, was proved.

The newborn child does not secrete tears when crying, for the nervous paths connecting the psychical reaction with the lacrimal centre only develop after some weeks. As a rule, however, the reflex secretion of tears functions from birth. We find in a few older investigations (Kirschstein, 1894; Axenfeld, 1899) the information that, at least in certain cases, the reflex lacrimal secretion is lacking or is very little marked, just after birth. In this respect new researches seem to be necessary, but it does not seem quite absurd to agree with Sommer (1903) and to see the cause of at least some cases of congenital alacrimia in a failure to establish a connexion between the nervous pathways and the lacrimal centre, which produces an alacrimia congenita persistens. We may see an analogy between this and, for example, the congenital lack of fusion which in certain cases becomes permanent and is the cause of strabismus.

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