Severe Bilateral Upper Palpebral Coloboma and Bifid Nasal Tip.

*JOYA CHOWDHURY, F.R.C.S., F.R.C.S.E.**

CONGENITAL fissure or coloboma of the eyelids is a rare anomaly, and involves all the layers of the lid The commoner form of palpebral coloboma is an unilateral cleft of the lid; bilateral clefts occur more occasionally. Mustarde (1965) describes severe bilateral upper palpebral coloboma in a Mexican Indian child. The developmental origin of the eyelids is a complex one, and various theories exist regarding the formation of this defect. The theory of pressure from amniotic bands resulting in destruction or prevention of proper development of the eyelid, causing coloboma, has several supporters. Upper palpebral coloboma associated with other occulo–auricular findings have been described by various authors. In almost every case the anomalies described were restricted to the head. Such cases are often combined with epibulbar tumours, and support the theory, that they are the effect of locally acting external causes, Gaps in the eyebrow, with a hairy process of the scalp extending across the forehead to the gap, may be explained by pressure from amnion bands.

Ida Mann (1957) describes the eyelids developing as mesoblastic epithelial covered folds, meeting at the palpebral fissure to fuse. Differentiation of the palpebral structures occur during the stage of fusion. The epithelial adhesion breaks down by the end of the fifth month. Failure of adhesion of the palpebral folds or localised disruption of the adhesion may cause the anomaly. Various other facial anomalies such as macrostoma, facial clefts, cleft lip, accessory auricular appendices have been reported in cases having palpebral coloboma. Such combination of several anomalies is usually considered to support a heritable nature of the condition. However, in this instance the theory of heritance seems less applicable, as there are few reports to substantiate such a theory; and the defects are restricted to the face.

Kiskadden and McGregor (1947) observed fraternal twins, discordant as regards bilaterally symmetrical coloboma of of the upper eyelids.

Case Report :

Severe bilateral upper palpebral coloboma were seen in A.S. a boy aged four years. (Figure 1) The parents were not interrelated. The mother's pregnancy had been uneventful. There was no family history of congenital anomalies; and his two siblings, a girl aged twelve years and a boy aged one year, were normal. A.S. had been reared as a blind child, greatly loved by his family, and was very cheerful. His

*Calcutta Medical College Hospitals and Calcutta Medical Research Institute.

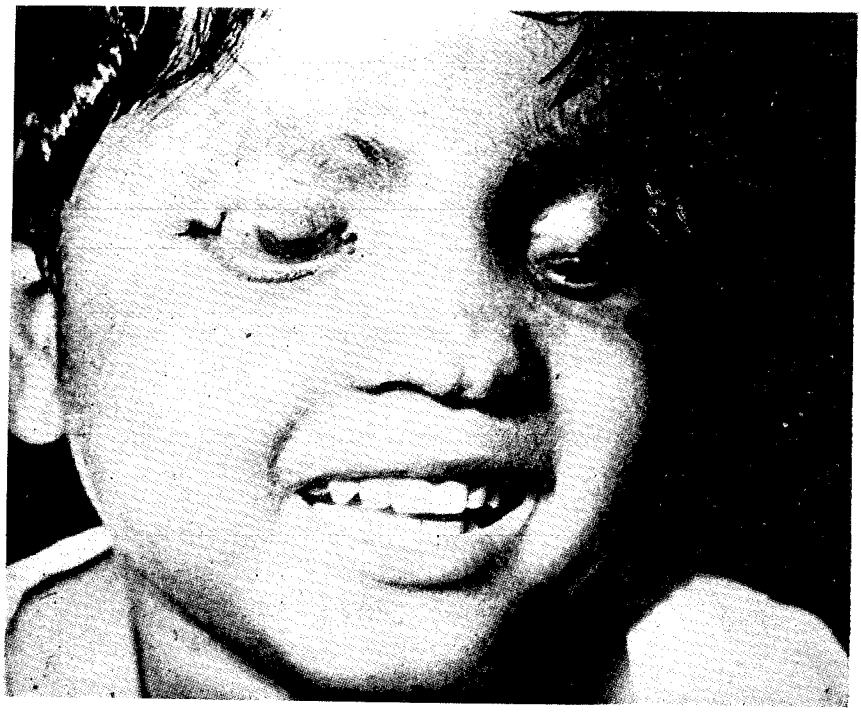

Fig. 1

parents had not sought any medical opinion previously, and he had no prophylactic treatment to prevent corneal damage. The coloboma in both eyes were similar in extent and involved practically the whole width of the eyelid. Minimal lid substance was present at the extreme ends, near the canthi, and carried a few lashes. A skin-like adhesion extended on to the eyeball in between the ends of the fissure and was adherent to or replaced, the upper two thirds of the cornea. The visible lower one third of the cornea was opaque. There was no light perception. The eyebrows were poorly defined and the forehead was hairy. The child also had separation of the alar cartilages and a bifid nasal tip. He had no other congenital anomaly.

Summary :

A case of severe bilateral congenital coloboma of the upper eyelids, also having bifid nasal tip is recorded.

References

1. Kiskadden W.S. & McGregor, M. W.: Coloboma of the Eyelids, Plast. & Reconst. Surg , 2 : 60, 1947.
2. Mann I. : Developmental Abnormalities of the Eye. Philadelphia, Pennsylvania, Montreal, Canada : M.B. Lipincott Co,, 1957.
3. Mustarde J. C. : Repair and Reconstruction in the Orbital Region". E & S. Livingstone Ltd., Edinburgh and London, 1966.