CONGENITAL EXTERNAL OPHTHALMOPLÉGIA*

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CONGENITAL external ophthalmoplegia can occur in two forms, complete and partial. Duke-Elder (1964) states that complete external ophthalmoplegia is rare as a congenital condition, but a number of cases are found in the literature where the eyes have been described as immobile. Partial external ophthalmoplegia occurs in all grades and varieties, from a very slight movement or only a tremor on effort (Cooper, 1910; Bradburne, 1912) to limited movements in all directions (Giri, 1936). In the present case both the eyes were immobile except for a very slight jerk to the right in the right eye and to the left in the left eye when the patient tried to look at a finger moved in these directions.

Case Report

A Hindu boy aged 12 years was brought to the ophthalmic out-patient department of the Jai Arogya Hospital, Gwalior, with the complaint that he had never been able to open his eyes fully. His parents and his two brothers and one sister showed no abnormality. There was no family history of any congenital ocular anomaly.

On Examination.—The boy had a characteristic appearance. He kept his head slightly tilted backwards, with arching of both eyebrows from compensatory overaction of the occipito-frontalis muscle. Both the upper eyelids were ptosed, with smooth, unwrinkled, thin skin and without a tarsal fold. Only the lower half of the cornea was visible (Fig. 1). On pressing the brows firmly against the supra-orbital margin inability to raise the lids was complete.

On testing the movements of the eyeball by asking the patient to follow a finger held in front of him, the eyes were immobile, except for a very slight jerk of the right eye on looking right and of the left eye on looking to the left (Figs 2–5). The conjunctiva, cornea, anterior chamber, and iris were normal in both eyes. The pupils were normal in appearance and reacting in both eyes. Fundus examination showed no abnormality and visual acuity in both eyes was 6/12.

Treatment.—A Blascovitz operation for the correction of ptosis was performed in the right eye. The recovery was uneventful and there was satisfactory correction of the ptosis in the right eye (Fig. 6). During the operation the levator muscle and its aponeurosis were found to be normally developed and could be easily separated and resected. As the parents wished the operation on the left eye to be done at a later date the patient was discharged from hospital.

Discussion

Hereditary transmission of ophthalmoplegia is common, but in the present case there was no familial or hereditary incidence. The palsy is generally considered to be due to a fault of the central mechanism (a nuclear aplasia or hypoplasia). The

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FIG. 1.—The patient looking straight ahead.

FIG. 2.—Following the examiner's finger, looking right.

FIG. 3.—Looking left.

FIG. 4.—Looking up.

FIG. 5.—Looking down.

FIG. 6.—After correction of the ptosis of the right eye.

presence of a normally developed levator muscle in this case shows that it was due to a defect of the central mechanism, and lends further weight to this theory of origin.

Summary

A sporadic case of congenital external ophthalmoplegia with immobile eyes is presented. During a correcting operation for ptosis the levator muscle of the right eye was found to be normally developed, showing that the central mechanism was at fault in the present case.

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