VARIANT OF DUANE'S RETRACTION SYNDROME*

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THE retraction syndrome was first reported by Stilling (1887). Duane (1905) reported a series of 54 cases in detail and the condition has come to be known as Duane's retraction syndrome. It involves a congenital abnormality of the horizontally acting intra-ocular muscles, most commonly the lateral rectus, and is not associated with any lesion of the central nervous system. It is usually uniocular and appears to be more common in females. Characteristically there is complete limitation of abduction, with retraction of the globe and a marked upshoot or downdrift of the affected eye on attempted adduction.

It is agreed by most authors that the lesion is peripheral, and that the limitation of movement of the lateral rectus is due to the replacement of muscle tissue by an inelastic fibrous band (Duke-Elder, 1949). This fibrous band can neither contract nor relax as normal muscles do, so that on contraction of the medial rectus the eyeball is retracted backwards instead of moving normally inwards. Some workers attribute the retraction of the eyeball to faulty insertions of the medial rectus muscle and check ligaments, while others have postulated the presence of an atavistic retractor bulbi.

The fact that diplopia is usually absent is understandable, because the syndrome is present from birth.

In the Gandhi Eye Hospital, we have come across a number of cases with the usual features of Duane's syndrome, but the case reported below presented certain unusual features, including diplopia.

Case Report

A girl aged 16 years had had diplopia and strabismus in the right eye since childhood. She could overcome the diplopia by turning the head to the right.

Visual Acuity.—6/6 with glasses in both eyes.

Head Posture.—The head was turned to the right and tilted to the left shoulder.

Cover Test.—The right eye showed excessive divergence and slight elevation, especially on any attempt to adduct the left eye (Fig. 1a). The left eye was normal.

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Ocular Movements.—There was no deviation in the primary position (Fig. 1b).

Unocular Right eye, normal.
Left eye, abduction absent beyond the midline.
Adduction limited, and when attempted caused retraction of the eye-ball and narrowing of the palpebral fissure.

FIG. 1(a).—Position adopted by right eye on cover test.

FIG. 1(b).—Primary position with forcible straightening of head.

FIG. 1(c).—Dextroversion.

FIG. 1(d).—Laeoversion.

Binocular Dextroversion: Updrift of right eye, accompanied by narrowing of the palpebral fissure and enophthalmos of the left eye (Fig. 1c).
Laeoversion: Excessive adduction of right eye and limitation of abduction of left eye with widening of the palpebral fissure (Fig. 1d).

Convergence Right eye, normal.
Left eye, very poor.

Hess Screen Test.—Paresis of left medial rectus and overaction of right lateral rectus (Fig. 2).

FIG. 2.—Hess screen test.

Diplopia Test.—Crossed diplopia to the right and a variable amount of homonymous diplopia to the left.

Binocular Field of Fixation.—Diplopia in the whole binocular field except for a small area in the left-hand lower quadrant.

This case showed the following unusual features:

- (i) The eyes were straight in the primary position, with involvement of both lateral and medial recti of the left eye.
- (ii) Although there was a marked limitation of movement of the left lateral rectus, there were all the signs of a paralysis of the left medial rectus: compensatory head posture, Hess screen test, diplopia test, and binocular field of fixation. When the head was forcibly straightened, diplopia resulted.
- (iii) The overaction of the right lateral rectus as revealed by the cover test further emphasized the apparent paralysis of the left medial rectus (Fig. 1a). There was also over-action of the right medial rectus on laevoversion, as expected.

Summary

This case has been described in some detail because variants of the classic syndrome pose certain problems in diagnosis and treatment.

It appears that the left medial rectus muscle either has a gross anomaly of insertion or is fibrotic in nature, so masking the effect of the left lateral rectus palsy. The exact nature of the defect could only be established by exploratory operation, to which the patient would not agree.

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