Yonsei Medical Journal
Vol. 41, No. 1, pp. 155−158, 2000

A Case of Congenital Inverse Duane's Retraction Syndrome

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Abstract

Inverse Duane's retraction syndrome is very uncommon. Congenital cases are even more unusual. A 6-year-old girl with convergent squint along with severe restriction on abduction is described. On attempted abduction, a narrowing of the palpebral fissure, upshoot and retraction of the eyeball were observed. Brain and orbit MRI demonstrated no intracranial or intraorbital mass, fracture, or entrapment of the medial rectus. Forced duction test was strongly positive. The primary lesion was found to be a tight medial rectus with shortening and soft tissue contracture. Surgical tenotomy of the medial rectus led to successful postoperative motility, but some limitation at full adduction and abduction persisted. This is a case reported with congenital medial rectus shortening, suggesting that this condition may be one of the etiologies of the rare inverse Duane's retraction syndrome.

Key Words: Forced duction test, inverse Duane's retraction syndrome, tenotomy, tight contracture

INTRODUCTION

Duane's retraction syndrome is a well-known congenital musculo-facial anomaly.[1] The etiology of the syndrome is found to be complex. The electromyographic data suggest that there is a paradoxical innervation in Duane's retraction syndrome.[2] Co-contraction of the medial and lateral rectus muscles in adduction was thought to be responsible for the retraction of the globe with the explanation that the oculomotor nerve may send fibers to the lateral rectus.[3] There has been another suggestion that most of the innervation to the extraocular muscles reaches them via the gamma efferent fibers. These carry impulses to the muscle spindles. The spindles function as length setters or automatic load compensators. They act via the servo-loop through the spindle afferents and the main alpha motor neurons. There exists a direct anatomical link within the muscle by which the alpha motor neurons influence the discharge of the spindle afferents. Should this link be disturbed by the muscle being abnormally stiff, then this servo-loop would be broken or drastically modified. As a result, much of the innervation destined for the alpha motor neurons and the main muscle fibers would never reach them.[4]

Inverse Duane's retraction syndrome is a condition with reverse clinical features.[5] Abduction of the affected eye is possible to some extent and it is accompanied by retraction of the eyeball, narrowing of the palpebral fissure, and pseudoptosis. There may be some retraction of movement on adduction. The primary lesion is suspected to be the medial rectus muscle. Frequently, the muscle is of this sort following trauma to the medial wall of the orbit. In both conditions, with the limitation of abduction, the patient generally shows an esotropia in the primary position. There is only one case of congenital inverse Duane's retraction syndrome reported by Chatterjee et al., of which the cause was found to be extensive fibrous bands surrounding the medial rectus muscles extended to the medial orbital wall.[6] However, we report a typical case of congenital inverse Duane's retraction syndrome whose medial rectus muscle showed no entrapment from trauma or medial orbital wall-related deformity.

CASE REPORT

A 6-year-old girl had complained of her squint

Received July 14, 1999
Accepted September 3, 1999
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However, this congenital case showed amblyopia suggesting other characteristics of congenital inverse Duane's retraction syndrome. Shortening of the medial rectus muscle is suspected to be the main etiology of the congenital inverse Duane's retraction syndrome.

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