Longstanding Brown Syndrome : An atypical presentation and its management

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Abstract

Brown syndrome is an ocular motility disorder characterized by limited elevation in adduction. The congenital form of Brown syndrome is thought to occur due to anomalies of the superior oblique (SO) tendon or of the trochlea. The diagnosis of Brown syndrome is essentially clinical. The limitation in adduction and the positive forced duction test are the consistent features to diagnose Brown's syndrome. This patient exhibits, limitation of elevation on adduction that improved in abduction; widening of the palpebral fissure on adduction; and a positive forced duction test. The atypical features that made this case unique are a large hypertropia in the contralateral eye with a large constant exotropia and absence of AHP. It can be explained that congenital ocular motility disorders if not assessed and surgically corrected at younger age may cause variable symptoms depending on the severity. Bilateral LR recession and SO tenotomy in the right eye was performed, achieving satisfactory alignment with one surgical procedure.

Introduction

Congenital form of strabismus is often identified in early childhood and usually present with features like constant deviations or abnormal head postures (AHP). But when they present late, these presentations may vary among patients. This paper is to report a longstanding case of congenital Brown syndrome affecting one eye with uncommon presenting features.

Brown syndrome is an ocular motility disorder characterized by limited elevation in adduction. [1] Harold W. Brown first described the disorder in 1950 and initially named it the "superior oblique tendon sheath syndrome". The syndrome may be congenital or acquired. The congenital form of Brown syndrome is thought to occur due to anomalies of the superior oblique (SO) tendon or of the trochlea. [2, 5] The causes of the acquired form, are trauma, neoplasm, or systemic inflammatory conditions such as rheumatoid arthritis and systemic lupus erythematosus.

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The common clinical features of Brown syndrome are;

- Restricted forced ductions to elevations in adduction with near normal elevation in abduction;
- Same amount of limitation with version and duction; a divergence pattern in straight-up gaze;
- Widening of the palpebral fissure on adduction;
- Compensatory chin up position. [3,4]

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The less common clinical features are

- A mild elevation deficit or hypotropia in primary position,
- Downshoot in adduction. [5]

The diagnosis of Brown syndrome is essentially clinical. The finding of an eye that does not elevate on moving inward but elevates on moving outward with aligned eyes in primary gaze strongly suggests Brown syndrome.

Exaggerated Forced duction testing may be necessary to confirm the diagnosis. CT and Magnetic resonance imaging (MRI) are also an essential tool for the diagnosis and management of congenital and acquired Brown syndrome.

In most of the publications presenting features of Browns syndrome were vertical misalignment and abnormal head posture. However, some reported associated horizontal deviations also. [5] But very few described features related to the unaffected eye in unilateral cases. The current reported Brown syndrome case is atypical in presentation, with a large contralateral hypertropia and exotropia.

Case Presentation

An eighteen years old male presented with complain of large upward and outward deviation of the right eye since early childhood. As no photo of childhood was available, the severity of the condition could not be documented. There is no history of diplopia or eye and head trauma. His birth, developmental and medical history was insignificant with no family history of strabismus.

His unaided visual acuity was 6/12 in the right eye and 6/6 in the left eye. After refraction, his visual acuity improved to 6/6p in right eye with -1.00 DS/- 4.50 DC at 180 degree and 6/6 in left eye with -1.00DC at 180 degree. On examination he had no abnormal head posture (figure-1). The cover test showed

large right hypertropia (RHT) and exotropia with left eye preference for both distance and near. When fixing with right eye there was left hypotropia and exotropia with left pseudoptosis (figure-2). Binocular single vision (BSV) and stereopsis were absent. Ocular motility and prism cover test measurements are shown in figure -3.

Figure-1 No AHP

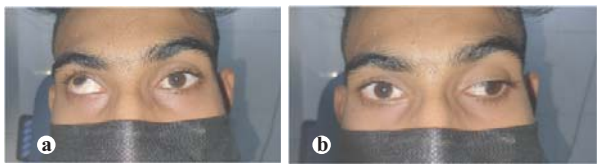

Figure 2 (a) Right hypertropia on fixing with left eye (b) Left hypotropia with pseudoptosis when fixing with right eye

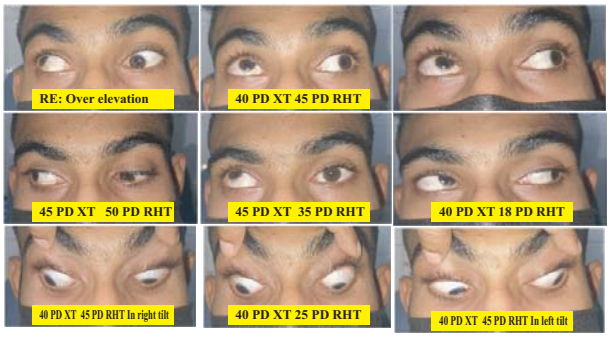

Figure -3: Nine gaze pictures with prism cover test measurements

The left eye showed limited elevation of -4 on adduction that improved to -2 in abduction both in duction and versions. The right showed +2 IOOA in adduction and over-elevation of +3 on dextroelevation. Mild widening of lid fissure was found in left eye during adduction, which was not present in the right eye.

Fundus photograph showed + 2 degree of

intorsion in left eye and no torsional abnormality in right eye(figure-4), suggesting inferior oblique palsy of left eye. But the Park's three-step test revealed normal finding, thus ruling out isolated inferior oblique muscle palsy of left eye. Right hypertropia in dextroversion is larger than in levoversion indicates Brown syndrome of left eye. No pattern was present.

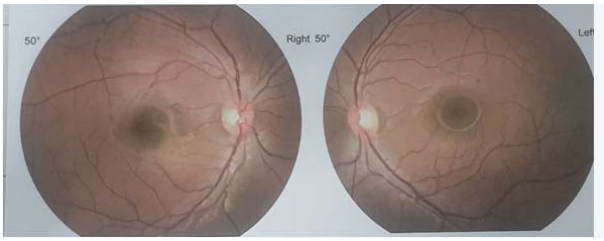

Figure-4: Mild intorsion of left eye

Forced duction test (FDT) and forced generation test (FGT) performed in the clinic, revealed no gross muscle tightness with normal muscle function.

Intraoperative FDT was performed, and a tight superior oblique in left eye confirmed the diagnosis of Brown syndrome. The picture of FDT done before and after performing superior oblique tenotomy is shown in figure -5.

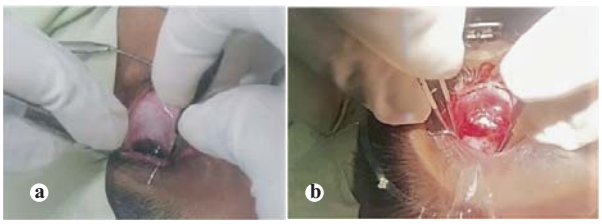

Figure-5: (a) FDT showing restriction due to tight SO (b) FDT showing release of restriction after SO tenotomy

On examining the Superior oblique tendon, the insertion was found more posterior than its normal position with no abnormal band or fiber. A complete Superior oblique tenotomy was performed in the left eye and bilateral 8.5mm lateral rectus recession was performed to correct the exodeviation in primary position. The surgery was performed through limbal incision in both eyes and no complications were noted. Postoperatively, the exotropia reduced to 6PD for distance and near. There was residual left hypotropia of 15PD in primary position (Figure-6). A remarkable improvement of elevation in adduction in the left eye was observed. BSV and stereopsis were absent.

Figure-6: Post-operative three months showing complete correction of exotropia with residual left Hypotropia of 15pd

The patient was followed up 6 months after surgery and on orthoptic evaluation there was no horizontal deviation for both distance and near with left hypotropia of 8 PD in primary position.

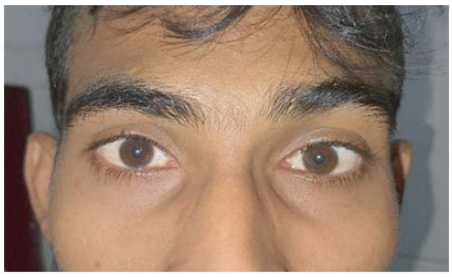

Figure-7: Post operative six months showing complete correction of exotropia with left Hypotropia of 8PD

Discussion

The clinical features initially described by Brown, for this ocular motility disorder were supported by several other studies. [3,7] Among these criteria the limitation in adduction and the positive forced duction test are the consistent features to diagnose Brown's syndrome. This patient exhibits limitation of elevation on adduction that improved in abduction; widening of the palpebral fissure on adduction; and a positive forced duction test. The atypical features that made this case unique are a

large hypertropia in the contralateral eye with a large constant exotropia and absence of AHP. However, horizontal misalignment associated with brown syndrome have been reported in several studies. The incidence noted was 18%- 86% [5,9], and exotropia was found more frequently than esotropia[8,9].

The main differential diagnosis that should be considered in this patient is an isolated left inferior oblique paralysis [9]. This very rare condition mimics Brown Syndrome, in the limitation of elevation in adduction. The positive forced duction test, however, is the major distinguishing feature. [10] Inferior oblique paralysis is mainly characterized by a positive Park's Three-step test and negative forced duction test. In addition, there is usually associated with an A pattern and overaction of the superior oblique muscle, which were absent in this patient. Another differential diagnosis is double elevator palsy. This condition, however, is associated with limitation of elevation on abduction that is equal to, or greater than, that on adduction, unlike in Brown syndrome and is also associated with ptosis of the eyelid.[6] The presence of a history of squint since childhood appears to be indicative of a congenital form of Brown syndrome in this patient. The presence of large hypertropia and suppression of the right eye are suggestive of a longstanding disturbance of the visual inputs during the period of visual development.

Unilateral Brown syndrome associated with contralateral large hypertropia has rarely been reported and there is little information on clinical findings of the unaffected eye. Contralateral IOOA however, has been reported to be associated with unilateral BS. [5] The reason for this large hypertropia in the right eye could be due to overaction of both the elevators of that eye, superior rectus and inferior oblique. Here, the patient's preferred eye is the left eye, which is the better seeing eye, but also the BS eye. Since it is a longstanding case that has no AHP, in an attempt to keep the left eye in primary position, the ipsilateral superior rectus exerts more force to overcome the restriction. This results in an increased innervation to the contralateral inferior oblique muscle based on the Hering's law, which postulates that both eyes receive equal innervation. The affected eye is limited, but the contralateral eye over elevates causing large hypertropia. The over elevation of the right eye on dextroelevation explains the overaction of superior rectus.

The surgical indication criteria for this case were the same as those described by Von Noorden [11]. Von Noorden's study proposed that surgery must be performed if there is hypotropia in the primary position, a significant anomalous head posture, and when Brown's syndrome is congenital and constant. Successfully SO tenotomy was proposed by Crawford in 1976 [12]. Several other procedures have been described to improve Brown's syndrome. Later, Von Noorden and others demonstrated that a SO tenotomy is the most effective surgery for Brown's syndrome [11, 13].

Conclusion

This case report shows that large hypertropia in the unaffected eye can be a presenting feature with unilateral Brown syndrome. It can be assumed that the hypertropia found in this patient would have been less severe if the condition had been diagnosed earlier and managed accordingly. It can be explained that congenital ocular motility disorders if not assessed and surgically corrected at younger age may cause variable symptoms depending on the severity.

Bilateral LR recession and SO tenotomy in right was performed as the first step of management. These procedures were successful in complete correction of the exodeviation and vertical deviation by 6months postoperatively. There should be more reports done with the long term follow up to set a better surgical plan.

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