Case Report:
Idiopathic Bilateral Sequential Brown Syndrome

SAEED ALQAHTANI, M.D.
The Department of Ophthalmology, Faculty of Medicine, Najran University

Abstract

Objective: To report a rare case of idiopathic Brown syndrome in the left eye first, then followed by Brown syndrome in the right eye after free tenotomy and inferior oblique anteriorization of the left eye.

Case Report: A 5-year old girl presented with ocular misalignment. The prism cover test at distant fixation showed hypotropia of the left eye, and chin up position which was 25 prism diopters (PD) in forced primary gaze, while in near fixation 35 prism diopters (PD). Eye movement of the left eye was markedly limited on elevation in adduction with normal elevation in abduction and markedly depression in adduction. Forced duction test of the left eye showed restricted elevation in adduction. Six months after surgical correction by free superior oblique tenotomy and inferior oblique anteriorization of the left eye, the girl showed hypotropia of the right eye, which was 35 prism diopters (PD) in primary gaze and markedly limited on elevation in adduction and limitation in elevation. Computerized tomography scan of the orbits ruled out any evolving local pathology. The right eye was managed conservatively for 6 months. Then, 10mm right superior oblique spacer was done combined with posterior fibers tenotomy and removal of the spacer in the left eye. Postoperatively, the patient was able to fix both eyes centrally with no misalignment, reasonable extra-ocular muscle movements and excellent vision.

Conclusion: Cases of BS can be identified by positive forced duction test. Management of BS is a real challenge to ophthalmologists. Nevertheless, conservative management for several months may prove effective in some cases of BS among children.


Introduction

BROWN syndrome (BS) is an ocular motility disorder, whose patients are characterized by being unable to gaze upward beyond the horizontal level while adducting their eye [1]. It is due to an abnormality of the superior oblique tendon sheath complex [2]. It is believed to be secondary to restriction of the superior oblique muscle in the trochlea/tendon complex that causes a tethering of the muscle when the eye is adducted [1].

Forced duction test for patients with BS shows severe mechanical restriction on attempts to elevate the adducted eye with no limitations of elevation in abduction. Commonly, there is widening of the palpebral fissure with adduction, while over-action of the ipsilateral superior oblique muscle is usually absent [3].

BS syndrome is either “congenital”, due to an anomaly of the trochlea and/or superior oblique tendon, or “acquired”, due to inflammatory, autoimmune, metabolic diseases, infection, systemic disease or trauma. Several surgical procedures can lead to acquired BS. However, literature on acquired BS in children is scarce and it is quite uncommon to report a case of acquired BS after free tenotomy of contralateral eye [4].

Herein, we report, a rare case of a child who presented with idiopathic BS in the left eye followed by BS in the other eye that appeared after free tenotomy and inferior oblique anteriorization of the left eye.

Case Report

In January 2013, a 5-year-old girl presented to the “Eyes Hospital” in Dhahran, Saudi Arabia, with left ocular misalignment. Her parents mentioned that they noticed the persistent misalignment in her eyes and her abnormal head position for the past two weeks.
The girl underwent detailed history taking. She, and also her parents, denied any relevant history of infection or trauma or exposure to any precipitating or triggering factor that would explain that condition. General examination was unremarkable.

Ophthalmologic evaluation (including visual acuity, slit lamp examination, intraocular pressure, and fundus examination) revealed no abnormal findings and refraction was +1.5 PD in both eyes. The prism cover test at distant fixation showed hypotropia of the left eye and chin up position, 25 (PD) in forced primary gaze near fixation (35 PD). Eye movement of the left eye was markedly limited on elevation in adduction with normal elevation in abduction and markedly depression in adduction. The provisional diagnosis of this case was “Idiopathic” Brown Syndrome in the left eye.

The case was managed conservatively and was followed-up for 6 months. However, no progress in her condition was observed. So, the plan was to do spacer 10mm for left eye. However, one month later, there was still left hypotropia in primary position (25 PD) with markedly limited elevation in adduction in the left eye. So, “left free tenotomy” was performed.

After tenotomy, there was no hypotropia in primary position and elevation in adduction improved, but with inferior oblique over-action (+3). There was left hypertropia in primary position with right limitation in elevation in superior gaze and esotropia in down gaze (Fig. 1). So, two months later, “inferior oblique anteriorization” was performed for the left eye.

Fig. (1): Left hypertropia in primary position with right limitation in elevation in superior gaze and esotropia in down gaze.
The patient remained orthophoric in primary position, with no abnormal head position for about 6 months. However, she started to show large angle of hypertropia OS (secondary angle) around 35 PD with right superior rectus under-action (−2) and she was unable to elevate the right eye in primary position and in adduction. A prism cover test at distance fixation revealed 4 prism diopters (PD) of right hypotropia in the primary position, 20 PD hypotropia in left gaze, and orthophoria in right gaze. Movement of the right eye became markedly limited on elevation in adduction while elevation in abduction was normal. To rule out any evolving local pathology, computerized tomography scan of the orbits was done and the results were normal. Fundus examination showed intorsion of the right eye. Forced duction test of the right eye demonstrated restricted elevation in adduction. The diagnosis was Idiopathic Brown Syndrome in the right eye.

The latest condition was managed conservatively for 6 months, but the case remained to show the same findings. So, 10mm right superior oblique spacer was done combined with posterior fibers tenotomy and removal of the spacer in the left eye.

Currently, more than 6 months after the last surgery, the patient is able to fix centrally both eyes. There is no more misalignment with reasonable extra-ocular muscle movements and excellent vision.

Discussion

BS describes the inability of a patient to perform an upward gaze while the eye is adducted due to an abnormality of the superior oblique tendon sheath complex [2,5]. The limited up-gaze in adduction seen in Brown’s syndrome is thought to be due to an abnormality of the trochlear tendon complex [6].

Our case, a 5-year old girl, presented with left ocular misalignment. Thorough history taking and physical examination of the case did not reveal a cause for that. So, the diagnosis of this case was “Idiopathic” BS in the left eye.

In accordance with the present finding, Fineman and Calhoun [7] stated that most cases of acquired BS are usually idiopathic. Kraft et al., [8] reported that in their case series of 5 children with BS, none of them had any evidence of systemic illness or local orbital disease to explain an acquired BS.

Management of our case was conservative for the first 6 months. However, since there was no improvement in her condition, a 10-mm spacer was performed followed by a “left free tenotomy” one month later. Moreover, after further two months, inferior oblique anteriorization was performed for the left eye, after which the patient remained orthophoric in primary position, with no abnormal head position for about 6 months. Afterwards, the case developed postoperative BS in the right eye. Forced duction test demonstrated restricted elevation in adduction. CT scan of the orbit was done to rule out any evolving local pathology and the results were normal.

Management of our BS case has been started conservatively since there have been reports of spontaneous resolution occurring sometimes over a short period of few months [9]. The mechanism by which resolution occurs is unclear. Reduced restriction of passage of the superior oblique tendon through the trochlear with growth of the eye has been suggested [6].

These multiple conservative and operative management procedures for our case were described by Lee [10], who stated that management of BS cases is a challenging problem. Nevertheless, it may show spontaneous resolution and conservative management is successful in around 75% of cases.

Development of postoperative BS in the other eye was reported by several authors. Kraft et al., [8] reported that in 5 cases, BS developed in the second eye after surgery was performed on the first eye.

Bansal et al., [6] noted that the natural history of BS is poorly understood. The recognition of BS can be accomplished by clinical examination and confirming the diagnosis with a positive forced duction test [11].

The case was managed conservatively for 6 months. Then, 10mm right superior oblique spacer was done combined with posterior fibers tenotomy and removal of the spacer in the left eye. Postoperatively, the patient was able to fix both eyes centrally with no misalignment, reasonable extra-ocular muscle movements and excellent vision.

In conclusion, this case of bilateral idiopathic BS is the first to be reported in Saudi Arabia that manifested sequentially, first in the left eye and then in the right eye after performing tenotomy and inferior oblique anteriorization of the left eye for the left eye. Cases of BS can be identified by positive forced duction test. Management of BS is a real challenge to ophthalmologists. Nevertheless, conservative management for several months may prove effective in some cases of BS among children.
Acknowledgement:

Special thanks to Dr. Laila Jaddawi for the generous permission to report this case from her clinic.

References


