Case Report

Sequential Presentation of Bilateral Brown Syndrome: Report of a Case with an Interesting Imaging Finding

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Abstract

Purpose: To present a case of bilateral Brown syndrome who presented as a unilateral disease and then showed the disease in the fellow eye in an older age.

Methods: A 4-year-old girl presented with congenital Brown syndrome of the left eye and underwent a superior oblique weakening procedure in that eye, but then developed Brown syndrome in the right eye which required two more surgeries on the right eye to attain an acceptable alignment.

Results: Her orbital computed tomography scan revealed that the distance between the annulus of Zinn and trochlea (Z-T distance) was 41.2 mm in both sides, which was comparatively longer than her age- and sex-matched cases. This finding can suggest a possible mechanism of Brown syndrome development in some patients.

Conclusion: Bilateral Brown syndrome can present as a sequential disease, and its radiologic finding may be associated with increased Z-T distance.

Keywords: Annulus of Zinn-trochlea distance, Brown syndrome, Sequential Brown syndrome, Superior oblique weakening

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Introduction

Brown syndrome is clinically characterized by the limitation of elevation in adduction in the affected eye. The clinical manifestations can be of different and perplexing scenarios such as congenital, acquired, constant, or intermittent presentations.[1,2] The natural course of the Brown syndrome is also variable in different patients, including a spontaneous improvement or gradual deterioration over the years.[1,2]

Brown syndrome presents bilaterally in 10% of patients, and from this group, there are rare reports of sequential involvement.[3-6] Herein, the authors introduce a patient with Brown syndrome with a sequentially bilateral presentation and her remarkable orbital imaging findings.

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Case Report

A 4-year-old girl presented to our strabismus clinic for the evaluation of the eye deviation and abnormal head posture that was evident since her infancy. Her medical history was otherwise unremarkable. On examination, she had a central and steady fixation in each eye. The full cycloplegic refraction was +2.5–2.50 × 140º in the right eye and +3.5–2.00 × 20º in the left eye. Her motility tests were remarkable for 25 prism diopter (PD) hypotropia and 30 PD esotropia of the left eye. Elevation of the left eye was entirely limited in the adduction, but only a partial limitation was noted in the straight upgaze and abduction. Moreover, there was a significant overaction of the left superior oblique. The range of movements was

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full in the right eye [Figure 1]. She also had a right face turn of about 20°. The findings were consistent with a congenital Brown syndrome of the left eye. She was prescribed the full cyclorefraction, and the surgery option was discussed with the family. The family declined the surgery at that time, but she eventually underwent surgery at the age of 7 years. Her forced duction test under general anesthesia showed a severe restriction of the left superior oblique and normal for the other muscles that confirmed the diagnosis. The surgical procedures included a 10 mm longitudinal elongation of the left superior oblique tendon. The superior oblique tendon was spared and exposed through an incision at the supranasal quadrant. The tendon was then split in a Z-shaped fashion, and the two ends were connected using Mersilene 5-0 suture (Mersilene, Ethicon, INC., Somerville, NJ, USA). In addition, a 6 mm recession of the left medial rectus and 7 mm resection of the left lateral recuts were performed. The left hypotropia was subsequently resolved, and the left esotropia decreased to 5 PD.

One year later, she returned complaining of upward drifting the left eye. In her examination, she had a left hypertropia of 15 PD in the primary position that increased in the left gaze. The right eye showed a complete limitation of elevation in adduction and a partial limitation of elevation in straight upgaze and abduction. She also had a new face turn of 10° to the left [Figure 2]. The findings were consistent with the acquired Brown syndrome of the right eye that warranted a complete inflammatory workup and rheumatology consult. All the tests and evaluations were unremarkable. We also requested a computed tomography (CT) scan of the orbit and paranasal sinuses, which revealed that the distance between the annulus of Zinn and trochlea (Z-T distance) was 41.2 mm in both sides. CT scan was otherwise normal [Figure 3a].

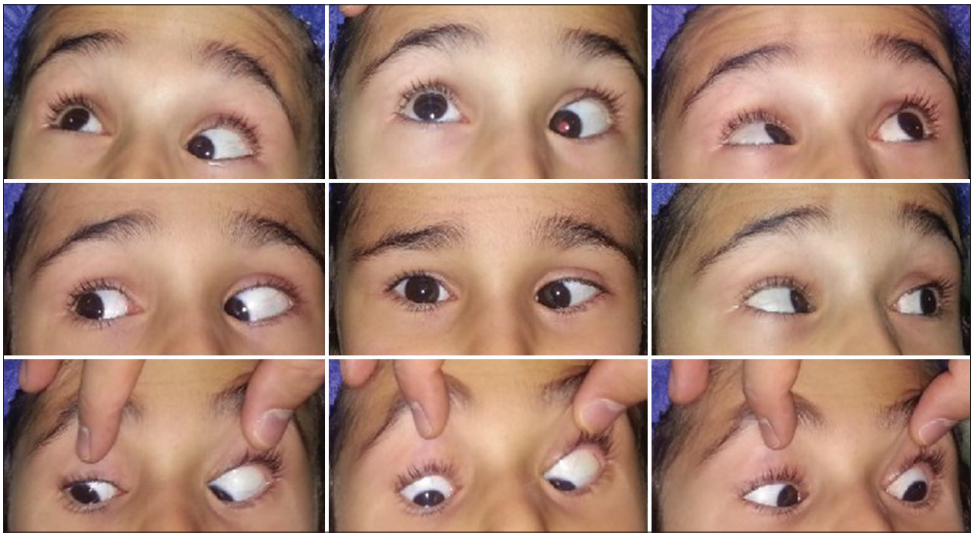

Figure 1: Clinical photographs of the patient's initial presentation. Left Brown syndrome was evident with esotropia, hypotropia, and limitation of elevation especially in adduction

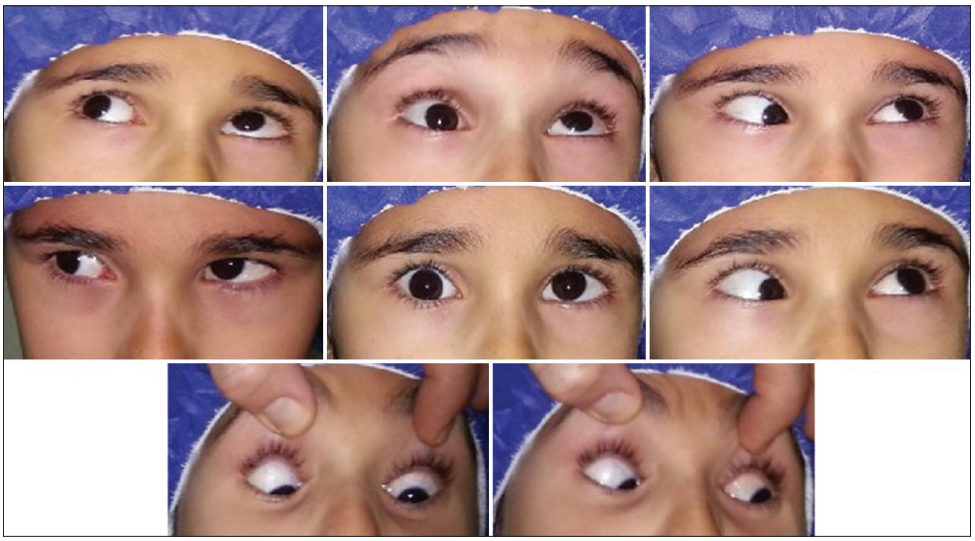

Figure 2: Clinical photographs of subsequent presentation of Brown syndrome in the right eye which was evident with hypertropia of the left eye and limitation of elevation especially in adduction in the right eye

The patient was monitored for the next 6 months without spontaneous improvement. Subsequently, a second surgery was planned. The forced duction test under general anesthesia confirmed a severe restriction of the right superior oblique tendon. We performed a longitudinal elongation of 10 mm on the right superior oblique tendon. This procedure did not significantly improve the deviation; thus, after 3 months, we performed a complete tenotomy of the right superior oblique at the nasal border of the superior rectus muscle. Subsequently, ocular movement limitations secondary to Brown syndrome resolved, and the head posture was corrected although she had a mild inferior oblique overaction and superior oblique underaction on the right side. In the primary position, she had a 6 PD left hyperphoria and 8 PD esophoria that was controlled with her anisometropic correction. Her follow-up over the next year showed stability in her alignment and eye movements [Figure 4].

Discussion

Although Brown syndrome presents bilaterally in 10% of patients, the sequential presentation has been rarely reported.[3-6] The time interval for the presentation of Brown syndrome in the second eye in prior reports ranged between 1 and 8 years,[3-6] as this interval was about 1 year in our patient. The Brown syndrome in our patient interestingly remained unilateral for 3 years since she was diagnosed, and she showed the signs of the Brown syndrome in the fellow eye only after the operation on the left eye.

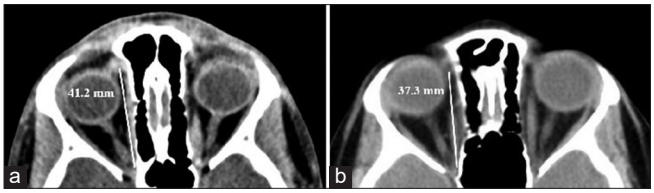

Figure 3: Axial computed tomography scan of the orbit, demonstrating the distance between annulus of Zinn and trochlea in our patient (a) and one representative age-and sex-matched control patient (b)

Kraft *et al*.[3] provided the largest series of sequentially bilateral Brown syndrome with 6 patients. In four cases, the severity of the involvement in the fellow eye advanced over time, which suggested a gradually evolving pathologic process. They performed bilateral superior oblique weakening in four patients of their series. Two patients remained under-corrected after the first surgery and underwent a second and more aggressive operation. Similarly, our patient required an augmenting superior oblique weakening procedure on the right eye.

The previous studies provided variable reports in terms of the severity of involvement in the fellow eye compared to the first eye.[3-6] In the series by Kraft *et al*.,[3] the clinical severity of disease in the fellow eye was equal to the first eye in 2/3 of cases as we observed in our patient. This may indicate a similar underlying mechanism in both eyes.

Although a structural abnormality may occasionally be found in the tendon-trochlea complex in acquired Brown syndrome, we did not find such abnormalities intraoperatively. Moreover, no inflammatory source for the acquired Brown syndrome was found in her systemic evaluations.

In an unpublished research, we measured the Z-T distance in 12 age- and sex-matched children who were admitted to the hospital and had a head CT scan for non-orbital reasons, such as head trauma. All non-contrast CT scans were performed using 64-slice scanner (Siemens sensation; Siemens Healthineers, Erlangen, Germany) in the supine position. For the purpose of reconstruction, 1 mm slice thickness and 1 mm reconstruction interval were used. All images were evaluated in picture archiving and communicating system by an expert radiologist in orbital imaging. The Z-T distance was measured in axial images in the medial aspect of both orbits at the level of maximum distance between annulus of

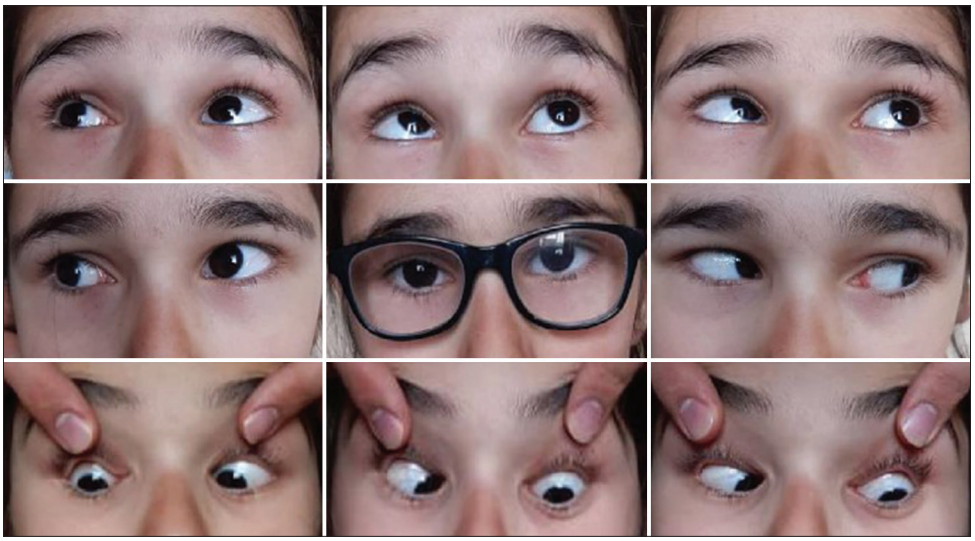

Figure 4: Clinical photographs at her last visit demonstrating a residual small angle hyperphoria and esophoria in the left eye. The limitation of elevation in adduction was substantially improved in both eyes

Zinn and trochlea. The average Z-T distance in this group was 37.1 ± 1 mm (range between 35.8 and 38.7 mm). In comparison, the child reported in this paper had a longer Z-T distance [Figure 3b].

Abrams[6] previously speculated the possible association between the elongated Z-T distance and development of the Brown syndrome in his report of a patient with sequentially bilateral Brown syndrome. The association between the short Z-T distance in plagiocephalic patients with apparent superior oblique palsy is a well-known phenomenon.[1,2] It seems that in an inverse mechanism, elongated Z-T distance and anterior displacement of the trochlea may impose traction on the reflected part of the superior oblique tendon. This imposed traction on the tendon inhibits its "telescoping" which presents as an acquired Brown syndrome.[6]

In conclusion, we presented a rare patient with the sequential presentation of bilateral Brown syndrome and discussed the management. We think that the clinical and radiographic findings in such patients can offer clues to understand the pathophysiology of this challenging disease better in future studies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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