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Unilateral Brown Syndrome Associated with Contralateral Superior Oblique Palsy in Two Brothers: A Case Report

Enam Danish[1], Ghaida Alahmadi[2], Abeer Aljahdali[3], Manal Hadrawi[4], Abdulelah Bana[5]

[1,4]*Department of Ophthalmology, King Fahd Armed Forces Hospital, Jeddah, Saudi Arabia.*
[2]*Department of Ophthalmology, Ohud hospital,Madina,Saudi Arabia.*
[3]*Department of Ophthalmology, Faculty of medicine, King Abdulaziz University, Jeddah, Saudi Arabia.*
[5]*Department of Radiology, King Fahd Armed Forces Hospital, Jeddah, Saudi Arabia.*

Abstract

The typical presentation of Brown syndrome is the restricted adduction on elevation and it still has a controversial pathology.We report a Cases of two brothers.The1st Case,8-year- old,had right head tilt with left hypertropia increase in right gaze and left head tilt.The right eye showed limitation of elevation in adduction.the2nd case,7-year-old had left head tilt with right hypertropia increased in left gaze and in right head tilt.The left eye showed limitation ofelevation in adduction.Magnetic resonance imaging of the orbits and brainstem showed hypoplasia of both SO muscles in case1 and hypoplastic left SO muscle with normal sized right SO muscle in case2.The trochlear nerve was not identified on either side in both cases.Congenital Brown syndrome associated with absent trochlear nerve and dysinnervationof the SO muscle or absent trochlear nerve with secondary changes of SO muscle that may manifest as Brown and SO palsy supported reports on congenital cranial dysinnervation disorders CCDDs as the underlying pathogenesis.

Keywords*: Brown syndrome, superior oblique palsy, superior oblique muscle, hypertropia , congenital cranial dysinnervation disorder.*

Introduction

The typical clinical presentation of a patient having Brown syndrome is the restricted elevation in adduction actively and passively during a forced duction test. Another feature is the minimal occurrence or absence of superior oblique overaction.[1,2] The head posture of individuals with identified Brown syndrome may even reveal a chin-up or head turn.[3] Mostof the cases are congenital and these are either autosomal recessive or autosomal dominant with reduced penetrance.[4]

Initial studies revealed that the congenital cases were related to superior oblique (SO) tendon sheath syndrome.[4] From recent evidence, a subtype of congenital Brown syndrome was suggested to belong to the spectrum of congenital cranial dysinnervation disorders (CCDDs). It is a group comprised of special forms of strabismus having a developmental defect of one or more cranial nerve nuclei, hypoplasia, or nerve deficit leading to abnormal innervation of the eye muscles. As a result, secondary abnormal structural changes were observed to the affected muscles as some patients had absent trochlear nerve with subsequent hypoplastic SOmuscle and other paradoxical innervation by the third cranial nerve.[5]

In this report, the unusual association of Brown syndrome with contralateral SO palsy was presented in the case of two brothers. To our knowledge, this is the first report of this case in our country.

Case Report

Two brothers were reported to have an abnormal head tilt for three years with no history of head/orbital trauma or ocular/orbital surgery. There had been a family history of head tiltwith the grandfather's cousin, and a previous medical record of the ocular examination was retrieved.

Case 1.The eight-year-old brother was noted to be healthy with normal growth and development besides the following observations. He was wearing glasses of -0.25 -1.25 X 2 right eye, -0.25 -3.00 X 180 left eye. His best-

corrected visual acuity was 20/25 OU. He also had a significant right head tilt (Fig. 1-A).

In the primary position, he had small exotropia (XT) of 10 prism diopters (Δ) with left hypertropia (LHT) of 20Δ on near and 12Δ on distance by prism cover test. He showed hypertropia 20Δ in right gaze, hypertropia 16Δ inleft gaze, hypertropia 12Δ in right head tilt, and hypertropia 24Δ in left head tilt. The righteye showed limitation of elevation in adduction (-2), and the left eye showed over elevation (+2) in adduction position (Fig. 1-B). No audible click or tenderness over trochlea in both eyes was remarked.

Maddox rod test could not be applied due to the difficulty of the test for the pediatric age group. The fundus photographs showed extorsion of the left fundus with no torsional abnormality in the right fundus (Fig. 3-A). Both eyes had normal optic nerve and macula. HR-MRI of the orbits in the coronal plane showed hypoplasia of both superior oblique (SO) muscles with the right muscle affected more than the left and The trochlear nerve was not identified on either side on the HR-MRI of the brainstem (Fig. 4-A). Therefore, leftinferior oblique myomectomy was planned for the patient.

At surgery, the forced traction test revealed a restricted elevation in adduction of the right eyethus confirming the diagnosis of true Brown syndrome. Three months after the surgery forthe follow-up, the head tilt was improved markedly (Fig. 1-C), and the orthoptic examination demonstrated a minimal flick of exotropia at near in the left eye. Both eyes also showed limitation of elevation in adduction (-2) in the right eye and (-1) in the left eye (Fig. 1-D).

Case 2. The seven-year-old brother, on the other hand, had rickets and asthma with which he was having a regular follow-up in the orthopedics and pediatric clinic. In addition, he was wearing glasses of +0.75 -1.25 X 169 right eye, +0.50 -1.00 X 15 left eye. His best-corrected visual acuity was 20/20 both eyes. He had a significant left head tilt with mild face turn (Fig. 2-A). In the primary position, he had small esotropia (ET) of 4 prism diopters (Δ) with right hypertropia (RHT) of 14Δ on near and 8Δ on distance by prism cover test. He showed hypertropia of 8Δ in right gaze, hypertropia of 20Δ in left gaze, hypertropia of 20Δin right head tilt, and hypertropia of 9Δ in left head tilt. The left eye showed limitation of elevation in adduction (-2) with a widening of the palpebral fissure, the right eye showedover elevation (+2) in adduction (Fig. 2-B). Also, no audible click or tenderness was observed over trochlea in both eyes.

Maddox rod test could not be applied due to the difficulty of the test for the pediatric age group. The fundus exam showed extorsion of the right fundus with no torsional abnormality in the left fundus (Fig. 3-B). For both eyes, there were noted normal optic nerve and macula. HR-MRI of the orbits in the coronal plane showed hypoplasia of the left SO muscle with normal-sized right SO muscle and The trochlear nerve was not identified on either side on the HR-MRI of the brainstem (Fig.4-B). Altogether, the observations and imaging results led to the right inferior oblique myomectomy of the patient.

At surgery, the diagnosis of Brown syndrome was confirmed upon the forced traction test, which revealed a restricted elevation in adduction of the left eye. During the follow-up, three months post-operatively, there was a significant improvement of the head tilt (Fig. 2-C). Additionally, the orthoptic examination demonstrated a straight eye at near and distance, and both eyes showed limitation of elevation in adduction (-1) in the right eye and (-2) in the left eye (Fig. 2-D).

Discussion

Described in this report is the unusual association of two siblings who developed a unilateral Brown syndrome with contralateral superior oblique (SO) palsy.

Two classifications have been established for Brown syndrome – true and simulated/pseudo. If the patient has the congenital form, the diagnosis is a true Brown syndrome while, the simulated or pseudo-Brown syndrome is the classification of those intermittent and acquired cases.[1] For our patients, the diagnosis made was true Brown syndrome because it wascongenital, and it was accompanied with restricted elevation in the eye adduction revealed from the positive traction test.

Four cases of absent trochlear nerve in congenital Brown syndrome were previously identified, wherein two had no bilateral trochlear nerve, and the other two had unidentified unilateral trochlear nerve on the side of Brown syndrome. In all the four patients, there wasno recorded muscle hypoplasia.[5] However, in the separate studies conducted by Ellis *et al*. and Yang *et al*., cases of Brown syndrome were associated with hypoplastic SO muscle.[6,7] In our report, case 1 showed bilateral hypoplastic SO muscle and no trochlear nerve. Thesecond case had hypoplastic left SO muscle with normal sized right SO muscle despite the absence of bilateral trochlear nerve that necessitates dysinnervation for the right SO muscle. The second evidence of right eye dysinnervation was the widening of the right palpebral fissure with elevation on adduction. This widening was explained to be because of the co- contraction of superior and inferior obliques pulling the eye outwards and retracting the eyelids as the SO and IO muscles are functional anterior to the eye equator.[8]

Ellis *et al*. proposed that congenital Brown syndrome could belong to the spectrum of CCDDas some patients had abnormal trochlear nerve development that might have led to the secondary changes in the SO muscle-tendon-trochlea complex.[6] This theory can explain the two cases described here, in which there was simultaneous occurrence of Brown syndrome inone eye and SO palsy in the other eye with confirmed abnormalities by HR-MRI.

Inferior oblique myectomy surgery was done for both cases and it showed dramatic improvement of the head tilt. The observations reported in the cases of the brothers strongly supported the association of congenital Brown syndrome with CCDD as one of its underlyingmechanisms especially for patients who had absent trochlear nerve and SO hypoplasia.However, the CCDD could not explain all the cases as spontaneous improvement occurred in 75% of the reports of congenital Brown syndrome.[9] Also, the presence of a positive forced duction test suggested that dysinnervation and tendon and/or trochlea anatomical pathologies may both responsible for the congenital Brown syndrome.

Conclusion

Association of congenital Brown syndrome with absent of the trochlear nerve with either changes in superior oblique muscle-tendon-trochlear complex or paradoxical innervation by third cranial nerve supported the CCDDs as the underlying pathogenesis. This is the first casereport in our region, and we recommend further studies to establish the pathogenesis of some cases of atypical congenital Brown syndrome.

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This case report did not receive any specific grant from funding agencies in the public,commercial, or not for- profit sectors.

Conflict of interest

The authors declared no potential conflicts of interest for this case report.

Ethical approval:

Permission from the ethical research committee in King Fahd Armed Forces Hospitalapproved this study.

Consent for publication

Informed written consent to publish data and images was obtained from the father.

Authors Contributions

ED and MH analyzed and interpreted the patient data .GA and AJ performed the examination, and were a major contributor in writing the manuscript. AB analyzedthe patient imaging. All authors read and approved the final manuscript.

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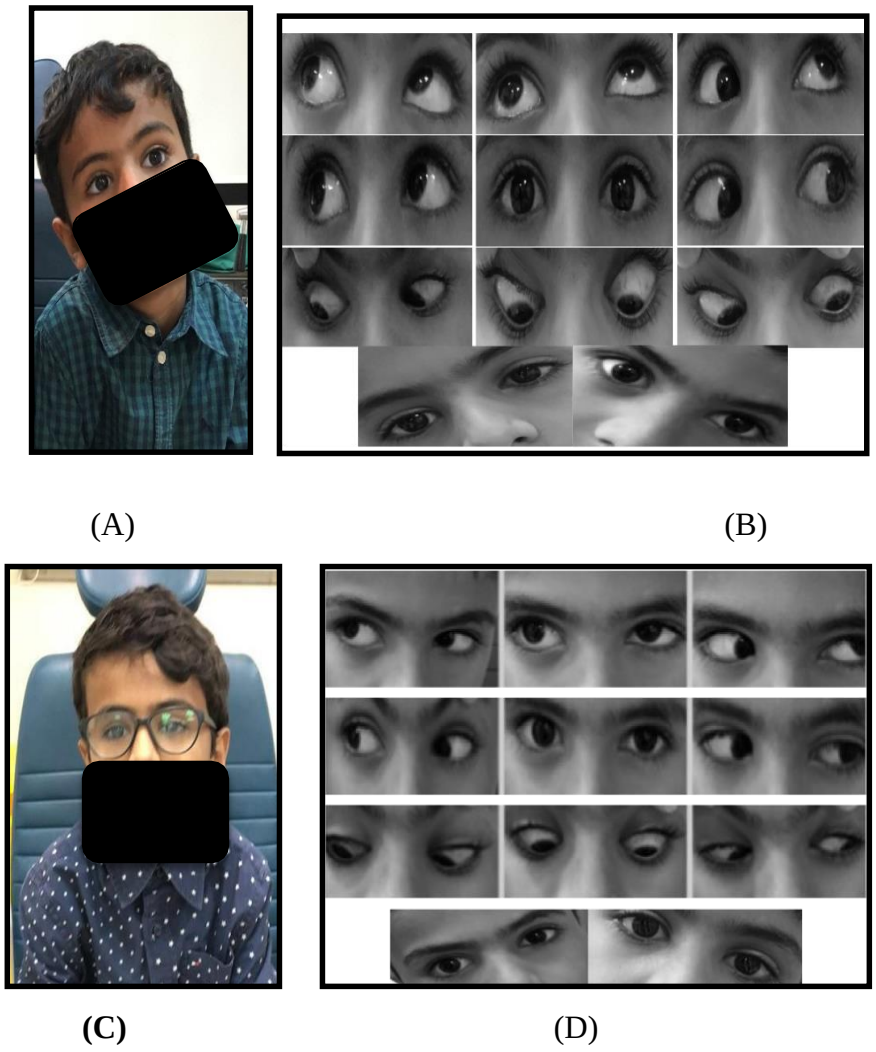
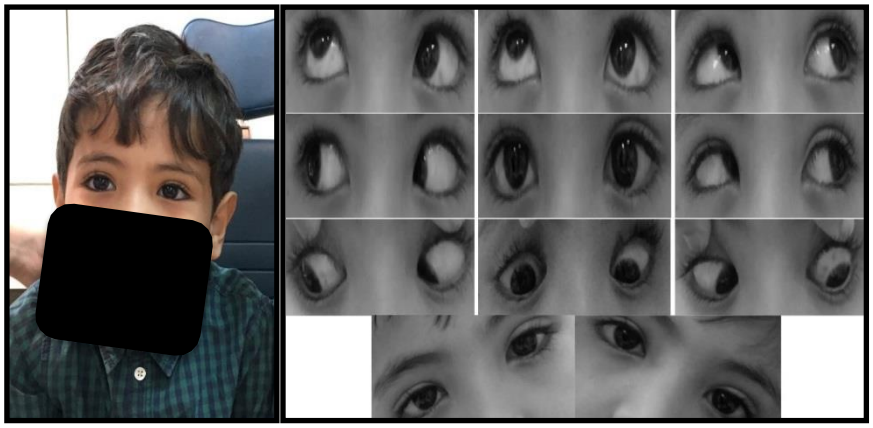


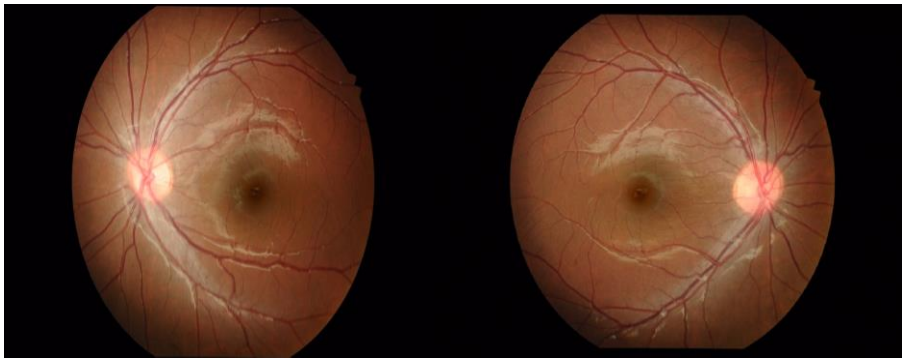
Figure 1 : (A) Preoperative photo showed right head tilt. **(B)** Preoperative Nine gaze positions and Bielschowsky head-tilt test showed limitation of elevation in adduction in the right eye and over elevation in adduction position in left eye, left hypertropia increase with right gaze and left head tilt. **(C)** Postoperative photo showed improvement of right head tilt. **(D)** Postoperative Nine gaze positions showed limitation of elevation in adduction in botheyes and No hypertropia in Bielschowsky head-tilt test.



(A) (B)

(C) (D)

Figure2:(A) Preoperative photo showed left head tilt. **(B)** Preoperative Nine gaze positions and Bielschowsky head-tilt test showed limitation of elevation in adduction in the left eye with a widening of the palpebral fissure and over elevation in adduction position in right eye,right hypertropia increase with left gaze and right head tilt. **(C)** Postoperative photo showed improvement of left head tilt. **(D)** Postoperative Nine gaze positions showed limitation of elevation in adduction in both eyes and No hypertropia in Bielschowsky head-tilt test.



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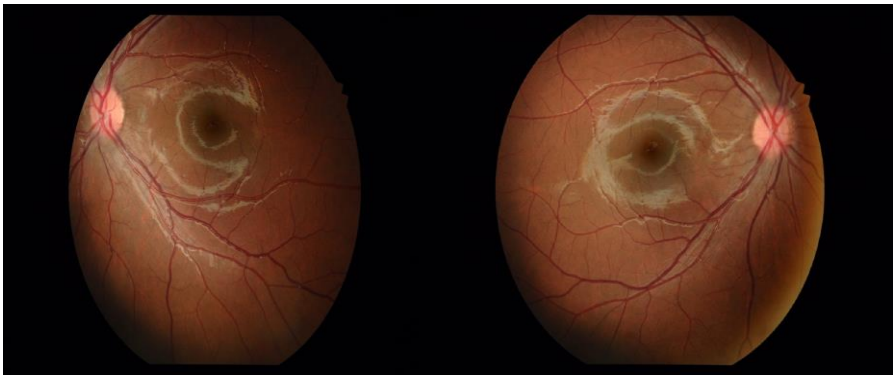
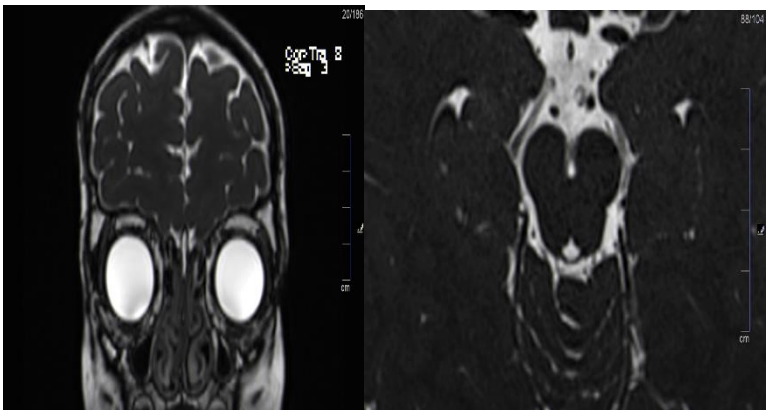


(B)

Figure3 : (A) The fundus photographs for the 1st case showed extorsion of the left funduswith no torsional abnormality in the right fundus. **(B)** The fundus photographs for the 2nd case showed extorsion of the right fundus with no torsional abnormality in the left fundus.



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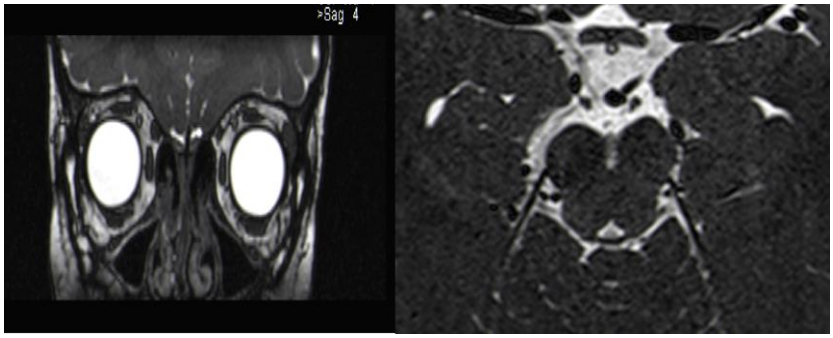

(B)

Figure4: (A) HR-MRI for the 1st case of the brainstem, the trochlear nerve was not observedon either side and HR-MRI of the orbits in the coronal plane showed hypoplasia of both superior oblique muscles with the right muscle affected more than the left**. (B)** HR-MRI for the 2nd case of the brainstem, the trochlear nerve was not observed on either side and HR- MRI of the orbits in the coronal plane showed hypoplasia of the left SO muscle