"Black curtain, Brown window" - A case of recurrent intermittent idiopathic Brown syndrome with atypical presenting symptoms

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ABSTRACT

Brown syndrome is characterized by limited elevation of the eye in an adducted position, most often secondary to mechanical restriction of the superior oblique tendon/trochlea complex. It can be constant or intermittent/recurrent.

We report on a 5 years-old boy who complained of seeing dark curtain and had strange head movements. He had similar symptoms 6 months prior. Examination revealed binocular vertical diplopia, multiplanar abnormal head position and limitation of the elevation of the right eye adduction only. Pain was elicited during attempt to elevate and adduct the right eye, with downshoot of right eye, and reflexive eyelid closure.

The patient was diagnosed with intermittent idiopathic Brown syndrome.

One day later, the patient was found to be asymptomatic, and had spontaneous resolution without any treatment.

Literature on acquired Brown’s syndrome in children is scarce. Very few have described the idiopathic intermittent or recurrent form. The acute, short and recurrent pattern of our patient’s symptoms showed an atypical, non-classical presentation and course. The initial misleading symptoms presented a diagnostic challenge. These might be explained by the prominent acute inflammatory component upon presentation, and we suggest two theories that might explain such an atypical clinical course.

1. Introduction

Brown syndrome is a vertical strabismus syndrome, characterized by limited elevation of the eye in an adducted position, most often secondary to mechanical restriction of the superior oblique tendon/trochlea complex. It was first described by Harold Whaley Brown in 1950. It accounts for approximately 2% of strabismus cases. Ten percent of cases are bilateral. There is no gender predilection.

Although it most often occurs as a congenital and constant condition, it can also be acquired, and some cases are intermittent, possibly recurrent. Most of these cases regress spontaneously.

We report on a child with atypical presentation of acute intermittent idiopathic Brown syndrome.

2. Case report

A 5 years-old boy presented to our pediatric emergency department complaining of several episodes of seeing a dark curtain in both eyes, which lasted a few seconds, and resolved spontaneously upon blinking few times. His mother also noticed some associated strange head movements and remembered that he had similar symptoms 6 months prior, with spontaneous resolution. Upon presentation, an abnormal head position was noted (Fig. 1) He strongly resisted any change in head position, and only when his head was straightened, he confirmed having a binocular vertical diplopia.

His past medical history included an idiopathic left sensorineural hearing loss diagnosed one year prior to admission and a cochlear implant surgery one week before presentation, with a normal post-operative course. There was no history of trauma. His family history was negative for ocular or neurological problems.

General physical examination revealed no abnormalities. Ophthalmic examination showed a multiplanar abnormal head position (Fig. 1): left face turn and left head tilt. Uncorrected visual acuity was 20/20 in both eyes. Pupillary reactions were normal. Tenderness over the right eye’s trochlear region was found, with no swelling or “click” sensation. The anterior segment was normal except for a right lower eyelid hordeolum. Funduscopic examination was normal.
Eye motility examination showed orthophoria in primary gaze position, and limitation of the elevation of the right eye that was most remarkable in adduction and normal in abduction (Fig. 2). Pain was elicited during attempt to elevate and adduct the right eye, and occasionally a downshoot right eye movement was noted (Fig. 3). The boy experienced an unpleasant feeling on every attempt to elevate the right eye on adduction, and it produced a reflexive closure of the eyelids (Fig. 4).

Blood tests including full blood count and serologic work-up for immunologic and inflammatory diseases such as Rheumatoid Arthritis, Systemic Lupus Erythematosus, and Sjogren syndrome, were inconclusive. A COVID-19 test was negative. Considering the boy’s age, it was decided not to perform a CT scan of the orbit or brain. MRI was contraindicated because of the cochlear implant. However, previous CT scan and MRI which were performed two weeks before presentation related to the cochlear surgery, demonstrated no ocular or orbital pathology.

A conservative approach was recommended, and no treatment was given.

One day later, the patient was found to be asymptomatic with a spontaneous resolution of the diplopia and abnormal head position. Ophthalmic examination findings were normal except of a small residual restriction of elevation of the right eye in adduction. The patient’s mother remembered a similar episode 6 months earlier which had resolved spontaneously within one day. There was not any documentation of the previous episode, apart from his mother’s description. A follow-up over 6 months was uneventful.

3. Discussion

We describe a young boy with an acute episode of restriction of elevation of the right eye in adduction, that resolved spontaneously over one day. The history suggests that this was a recurrent event. A specific cause of the temporary and short-lived eye movement restriction could not be identified.

Our patient’s restriction had mechanical characteristics. The combination of pain elicited on any attempt to elevate and adduct the eye, the tenderness over trochlear region, and the occasional downshoot movement of the right eye, was suggestive of a prominent inflammatory component, and an acute intermittent idiopathic Brown syndrome, or, considering the history, recurrent Brown syndrome. The literature on acquired Brown’s syndrome in children is scarce. Very few case reports have described the idiopathic intermittent form.

There are few theories regarding the cause of acquired idiopathic intermittent or recurrent Brown syndrome. The short course of the syndrome in our patient, combined with the prominent inflammatory component could be explained by the Bell’s phenomenon theory that was suggested in a Brown syndrome case report with a cyclic pattern. Most cases of acquired Brown syndrome are idiopathic and self-limited. However, it must be kept in mind that some cases are secondary to other pathologies, including inflammation, trauma, tendon cysts, previous sinusitis, orbital tumors, and iatrogenic causes such as orbital or strabismus surgery. These must be ruled out by a thorough history taking, physical examination, laboratory tests, and suitable imaging modalities.

Thus, the entity of Brown syndrome should be better recognized, and investigated.

Our case presented a diagnostic challenge. The boy’s complaints were not the classic “vertical diplopia”, but rather a sudden appearance of a ‘black curtain’, accompanied by some strange head movements. These might be explained by the prominent inflammatory component upon presentation.

The patient experienced such a strong and painful acute eye movement restriction, that made him close his eyes each time a painful movement was made, thus produced a black curtain appearance that lasted a few seconds until he blinked and “recovered”. His attempts to avoid the painful eye movements, made him turn his head instead of his eyes, to achieve the desirable gaze. Thus, producing strange head movements, with tilt and turn.

Our patient’s abnormal head position could have been attributed to his hearing problem, his recent cochlear implant surgery, or to a movement disorder (dystonia) such as oculogyric crisis. Only the eye motility examination revealed the true nature of his disorder.

The acute, short and recurrent pattern of our patient’s symptoms showed an atypical, non-classical presentation of Brown syndrome.

Two new theories may explain the recurrent pattern of the syndrome and its short course. We suggest the “Stretch theory” to explain the recurrent inflammatory pattern: when the eye is adducted, a congenitally short superior oblique tendon stretches. Repetitive stretching may cause friction between the tendon and its sheath, culminating in thickening of the sheath. A painful entrapment of the tendon may then lead to recurrent events of stenosing tenosynovitis or tendinitis. Furthermore, “the Avoidance theory” may explain the short course observed. The acute, strong pain causes the patient to avoid painful eye movements, lessening further tendon stretching and permitting quicker spontaneous resolution of the tenosynovitis, as in self-limited overuse injuries.

The management of such cases is based on a close follow up. A conservative approach is usually taken, as in our case.

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Fig. 1. Multiplanar abnormal head position: left face turn and left head tilt.
4. Conclusions

The child’s history of hearing loss with recent cochlear implant as well as his unique description of his symptoms which were misleading at first, presented a diagnostic challenge.

One should always consider ophtalmic or orbital pathologies, when dealing with strange head movements in children of any age, especially if accompanied by strange ophthalmic complaints.

Considering the possibility of such an atypical presentation, the entity of Brown syndrome should be better recognized, and investigated.

Patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient’s guardian (mother) has given her consent and granted permission for publication of the photographs that permit identification of the patient for educational purposes. The patient’s guardian understands that patient’s names and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Ethics approval and consent to participate

Not applicable.

Consent for publication

Was obtained and is attached to the manuscript.

Availability of data and material

Not applicable.

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Authors’ contributions

SV was first to examine the patient and suggesting diagnosis, analyzed and interpreted the patient data, and wrote the manuscript. RR and DR followed and treated the patient and was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

Declaration of competing interest

The authors declare that they have no competing interests.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2022.101378.

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