CASE REPORT:

A 5-YEAR-OLD BOY WITH ACUTE INTERMITTENT ACQUIRED BROWN'S SYNDROME

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ABSTRACT

We report a case of unilateral acute acquired intermittent Brown's syndrome in a 5-year old boy, that resolved spontaneously but recurred at regular intervals afterwards. A specific cause could not be found. We discuss the possible pathophysiological mechanisms of acquired Brown's syndrome in children.

KEYWORDS

acquired intermittent Brown's syndrome, acute diplopia, children

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Submitted: 20.04.2010
Accepted: 01.10.2010

INTRODUCTION

Brown's syndrome was first described by Brown in 1950 as "superior oblique tendon sheath syndrome" (1). He originally thought it was caused by secondary shortening of the anterior sheath of the superior oblique tendon from congenital palsy of the ipsilateral inferior oblique but this theory was later rejected as it became clear that pathology of the inferior oblique muscle does not play a role in the development of Brown's syndrome.

Brown's syndrome is characterized by a restrictive limitation of the elevation that is getting worse when the eye is in adduction. It can be classified as congenital or acquired, and may be constant or intermittent. Compared to the congenital form, acquired Brown's syndrome is more likely to be intermittent and to regress spontaneously (2).
This report describes a young boy with acute intermittent idiopathic Brown's syndrome of the right eye.

CASE REPORT

A 5-year-old boy first presented at our emergency department complaining of vertical diplopia after waking up in the morning. His vertical diplopia became more evident in upgaze. His mother noted a restriction of elevation of the right eye. He never experienced any eye problems in the past. There was no history of trauma or any medical illness. Family history was negative. Ophthalmic examination showed an uncorrected visual acuity of 9/10 in both eyes. Pupillary reactions were normal.

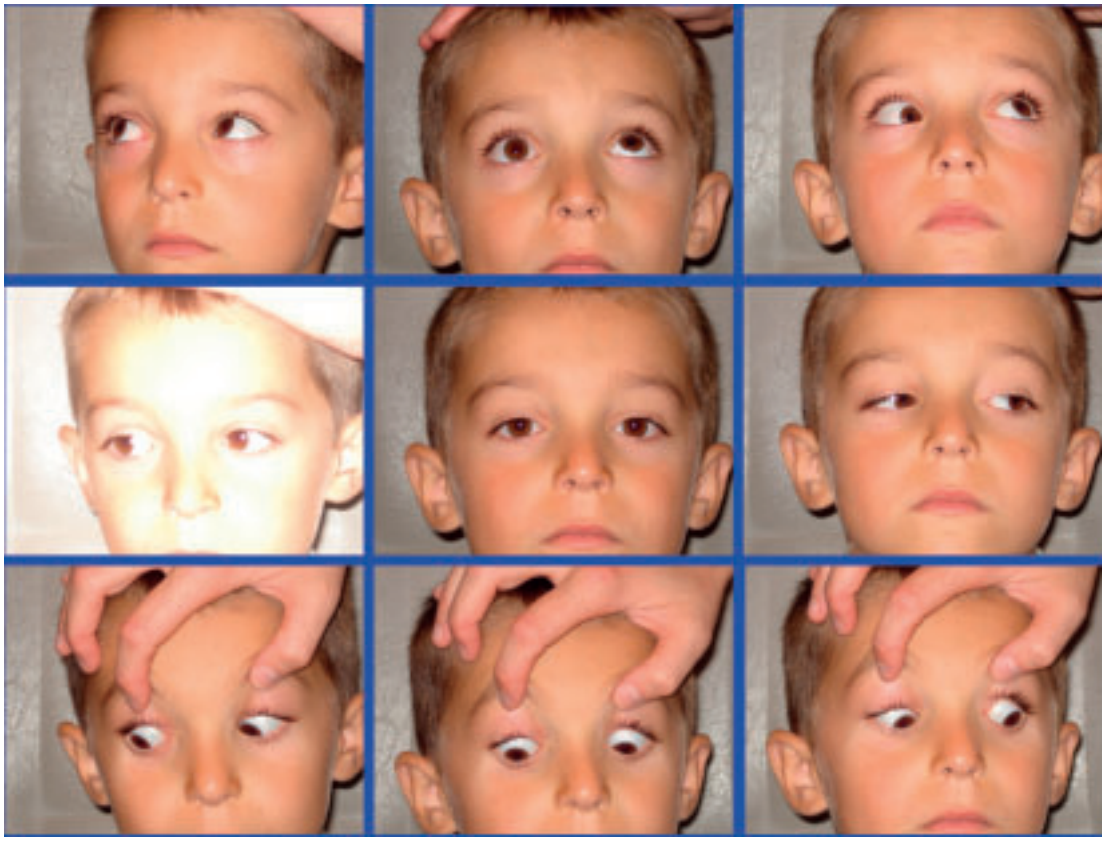

Fig. 1: Positions of gaze. Note the restricted elevation of the right eye when in adduction.

There was no swelling or tenderness in the trochlear region. The anterior segment and fundi were normal.

Motility examination revealed a limitation of the elevation of the right eye that was most remarkable in adduction, and normal in abduction. Eyes were orthophoric in primary gaze and there was no hypotropia present. Superior oblique overaction or A- and V-pattern were not present on examination. There was no abnormal head posturing or head tilt (Figure 1).

A Hess Screen Analysis was performed and confirmed the restriction. The same day, our patient was referred to a pediatrician for a thorough physical examination, which revealed no abnormalities. Blood tests including full blood count and serologic work-up for immunologic and inflammatory diseases were negative. A computed tomography scan (CT scan) of the orbit showed a symmetrical aspect of the eye musculature and the optic nerve. No intraconal or extraconal mass was seen. The superior oblique muscle tendon and the trochlea appeared to be slightly enhanced on the right side compared to the left side, with a normal aspect of the superior oblique muscle (Figure 2A, 2B). CT scanning of the brain was normal.

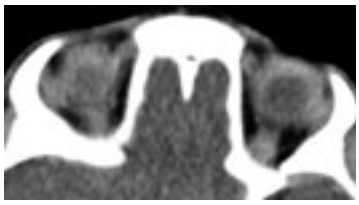

Fig. 2A: Axial orbital CT at the level of the trochlear/superior oblique tendon complex shows thickening of the insertion on the right side.

One week later, he was examined again and was found to be asymptomatic with a spontaneous resolution of the diplopia. Ophthalmic examination findings were normal except of a small residual res-

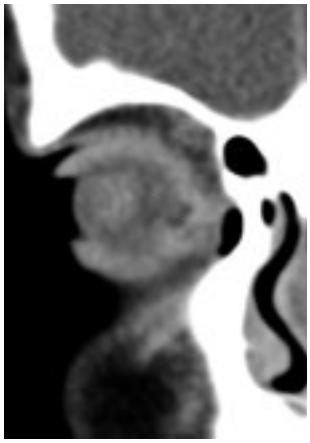

Fig. 2B: Coronal slice through the right eye shows the thickened trochlea/superior oblique tendon complex.

triction of elevation of the right eye in adduction (Figure 3).
Four weeks later, he had two other episodes of vertical diplopia and restrictive limitation of elevation in adduction of the right eye, with spontaneous resolution after two days.

Because of the intermittent character with spontaneous resolution, it was decided not to perform magnetic resonance imaging (MRI) of the orbit or brain or to repeat CT scanning.

DISCUSSION

The exact cause of acquired Brown's syndrome still remains controversial, but it is thought to be multifactorial. In adults, various pathological causes of Brown's syndrome have been described including traumatic, autoimmune, inflammatory, metabolic, neoplastic and infectious conditions, leading to a mechanical restriction (2). The literature on acquired Brown's syndrome in children is scarce. A few case reports have been documented of acquired Brown's syndrome in children with juvenile rheumatoid arthritis, pansinusitis, poststreptococcal reactive arthritis and hypogammaglobulinemia. Other described causes in children are superior oblique muscle cysticercosis, and trauma. Table I lists all described case reports of acquired Brown's syndrome in children.
In the majority of cases however acquired Brown's syndrome is idiopathic (2). Similarly, a specific cause of the temporary restriction could not be identified in our young patient.

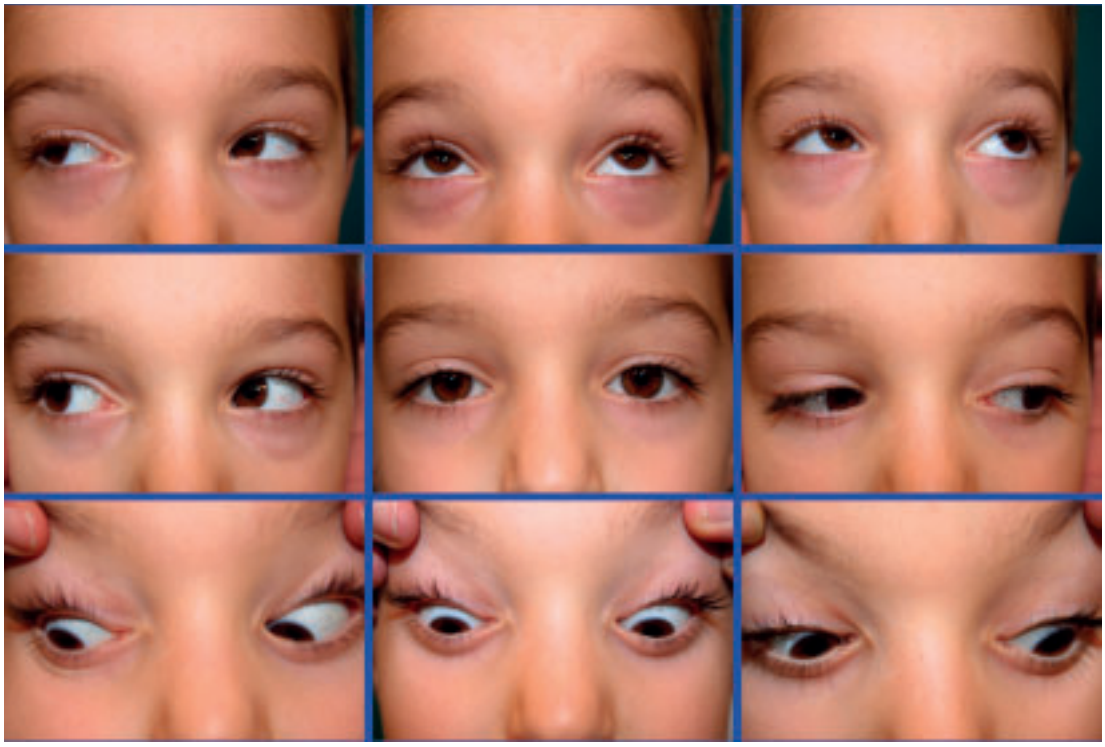

Fig. 3: Positions of gaze one week later. Note the complete resolution of the restricted elevation of the right eye in adduction.

Table I: *Articles about acquired Brown's syndrome in children.*

2009	*Abrams MS.*	*J Pediatr Ophthalmol Strabismus.* 2009 Mar-Apr;46(2):115-7.	23-month-old girl with progressive Brown's syndrome.
2004	Hargrove RN, Fleming JC, Kerr NC.	*J AAPOS*. 2004 Oct;8(5):507-8.	5-month-old girl with Brown'syndrome after surgical removal of hemangioma.
2002	Depras A, Chabrier S, Allard D, Gérard C, Lauras B.	*Arch Pediatr.* 2002 Jul;9(7):737-8.	10-year-old boy with idiopathic acquired Brown's syndrome.
2003	Rao VB, Sahare P, Varada V.	*J AAPOS*. 2003 Feb;7(1):23-7	Six patients with acquired Brown's syndrome caused by cysticercosis varying from 6-45 years (four patients younger than 18 years).
2001	Faust AO, Gillenwater JM, Saulsbury FT.	*J Rheumatol.* 2001 Dec;28(12):2748-9.	9-year-old boy with acquired Brown's syndrome and poststreptococcal reactive arthritis.
2001	Akar S, Söylev M, Onen F, Ada E, Birlik M, Akkoc N.	*Clin Exp Rheumatol.* 2001 May-Jun;19(3):354.	Child with juvenile idiopathic arthritis and acquired Brown's syndrome.
2000	Kraft SP, Nabi NU, Wilson ME et al.	*J AAPOS*. 2000 Jun;4(3):158-63.	Six patients (aged between 2-8 years) with unilateral congenital Brown's syndrome and delayed onset in the second eye.
1999	Wright KW.	*Trans Am Ophthalmol Soc.* 1999;97:1023-109.	Review article on Brown's syndrome in general.
1998	Siegel LM, DeSalles NL, Rosenbaum AL, Demer JL.	*Strabismus*. 1998 Mar; 6(1):19-29.	7-year-old boy with Brown's syndrome after dog bite.
1996	Brady KM, Hiles DA.	*Br J Ophthalmol*. 1996 Mar; 80(3):268-9.	2-year-old child with Brown's syndrome as a complication of resuscitation after fall in swimming pool.
1995	Can I, Yarangümeli A, Kural G.	*J Pediatr Ophthalmol Strabismus*. 1995 Jul-Aug; 32(4): 243-7.	15-year-old boy with acquired Brown's syndrome with cyclic characteristics.
1994	Bradshaw DJ, Bray VJ, Enzenauer RW et al.	*J Pediatr Ophthalmol Strabismus.* 1994; 31(2): 118-19.	Child with acquired Brown's syndrome associated with enteropathic arthropathy.
1991	Binkley K, Shore A, Buncic R, Roifman CM.	*J Rheumatol.* 1991 Jan; 18(1): 139-41.	10-year-old girl with hypogammaglobulinemia and progressive Brown's syndrome.
1990	Saunders RA, Stratas BA, Gordon RA, Holgate RC.	Arch Ophthalmol. 1990 Jan; 108(1):58-60.	5-year-old girl and 6-year-old boy with pansinusitis and acquired Brown's syndrome.
1989	Bradbury JA, Martin L, Strachan IM.	*Br J Ophthalmol*. 1989 Apr; 73(4):305-8.	5-year-old girl with acquired Brown's syndrome and mucopolysaccharosis.
1987	Baker RS, Conklin JD Jr.	*J Pediatr Ophthalmol Strabismus.* 1987 Jan-Feb; 24(1): 17-21.	Two children with acquired Brown's syndrome after trauma.
1985	Moore AT, Morin JD.	*J Pediatr Ophthalmol Strabismus.* 1985 Jan-Feb; 22(1): 26-30.	11-year-old boy with bilateral acquired Brown's syndrome and juvenile rheumatoid arthritis.
1984	Wang FM, Wertenbaker C, Behrens MM, Jacobs JC.	*Ophthalmology*. 1984 Jan; 91(1):23-6.	Two children with bilateral acquired Brown's syndrome and juvenile rheumatoid arthritis.
1984	Kemp AS, Searle C, Horne S.	*Ann Rheum Dis.* 1984; 43(5): 764-5.	9-year-old boy with transient Brown's syndrome and juvenile rheumatoid arthritis.
1974	Goldhammer Y, Smith JL.	*Neurology.* 1974 Jul; 24(7): 666-8.	11-year-old boy and 29-year-old woman with intermittent Brown's syndrome.

Despite the fact that in most patients acquired Brown's syndrome is of unknown etiology, it should be stated that a thorough examination with blood tests and imaging techniques is necessary in each patient presenting with acquired Brown's syndrome in order not to miss any localized or associated systemic disease. Especially CT and MRI can be helpful to identify possible mechanisms of Brown's syndrome, including trochlear damage, edema and tissue swelling, the superior oblique tendon abnormalities, abnormalities of rectus extra ocular muscle pulleys, and congenital abnormalities of superior oblique muscle (3).

In our case report, CT scanning showed only a slight enhancement in the area of the trochlea and the superior oblique tendon, suggesting a possible low-grade local inflammation, one theory regarding the cause of idiopathic Brown's syndrome (4). Swelling of the tendon within the trochlea would restrict tendon movement. Many of these acquired inflammatory Brown's syndrome cases result in intermittent Brown's syndrome and resolve spontaneously, as in our case. Helveston et al. suggested a telescoping mechanism to explain Brown's syndrome associated with local tendon inflammation. They showed that the elongation of the tendon is caused by movement of central tendon fibers and can be compared to the movement of a car antenna. According to these authors, constant or intermittent acquired Brown's syndrome might be caused by reduced telescoping elongation of the superior oblique tendon; excess fluid in the bursa-like structure within the trochlea could restrict tendon movement (2,5). Idiopathic inflammatory Brown's syndrome is often accompanied by the "click"-phenomenon and local superonasal pain or tenderness, even more by fever and edema. Especially in patients with a "click"-phenomenon, a trochlear massage or specific exercises can resolve the restriction of movement. Our patient however did not experience any of these symptoms.

Another suggested theory is the "wear and tear"-theory. Chronic movement of the superior oblique tendon through the trochlea can eventually lead to stenosing tenosynovitis, restricting the movement of the tendon through the trochlear tunnel (6). This theory has frequently been used to explain idiopathic acquired Brown's syndrome and is also called the Trigger Thumb Analogy Theory.

There is much discussion about the difference between congenital and acquired Brown's syndrome. Congenital Brown's syndrome was originally reported to be a constant restriction of movement that is present at birth, without pain or inflammation. A variety of congenital anomalies in the superior oblique muscle or the trochlear apparatus have been described (2). However it is postulated that some patients have an abnormal congenital insertion of the superior oblique tendon, but this may not become apparent until childhood after chronic movement of the tendon. These subtle forms of congenital Brown's syndrome that can intermittently become symptomatic can therefore be uncorrectly called acquired Brown's syndrome. Kraft et al. even reported six patients with congenital Brown's syndrome in one eye, who developed Brown's syndrome in the second eye without a clear underlying cause (7). This suggests that congenital and acquired Brown's syndrome are on a continuum with a common pathophysiology of restriction of free movements of the superior oblique tendon in the trochlea (2,7). According to this theory, it might be possible that a congenital anatomic anomaly in our patient predisposed the eye to reach a critical level of wear and tear and finally caused the intermittent vertical diplopia.

The fact that our patient experienced twice a restriction of the elevation of the right eye in adduction after waking up in the morning, could be explained by the fact that the superior oblique tendon is stretched with Bell's phenomenon during sleep, with possible fluid accumulation in the trochlear region. This mechanism is noted by Can et al., who reported a case of a 15-year old boy with acquired Brown's syndrome with a cyclic pattern, manifesting in the morning and resolving in the afternoon (8).

A conservative approach to management after a full investigation is justified in patients with idiopathic acquired Brown's syndrome since it resolves spontaneously in the majority of the cases, especially in intermittent forms (2,8). In our case, we also opted for a conservative approach.

REFERENCES

(1) Brown HW – Congenital structural muscle anomalies. Strabismus Ophthalmic Symposium, St Louis: CV Mosby, 1950; 205-36.
(2) Wright KW – Brown's Syndrome: diagnosis and management. Trans AM Ophthalmol Soc 1999; 97: 1023-109.
(3) Bhola R, Rosenbaum AL, Ortube, MC, Demer JL – High-Resolution Magnetic Resonance Imaging Demonstrates Varied Anatomic Abnormalities in Brown Syndrome. J AAPOS 2005; 9: 438-48.
(4) Hermann JS – Acquired Brown's Syndrome of inflammatory origin. Arch Ophthalmol 1978; 96: 1228-232.
(5) Helveston EM, Merriam WW, Ellfis FD et al. – The trochlea: A study of the anatomy and physiology. Ophthalmology 1982; 89: 124-144.
(6) Sanford-Smith JH – Superior oblique tendon sheath syndrome and its relationship to stenosing tenosynovitis. Br J Ophthalmol 1973; 57: 859-65.
(7) Kraft SP, Nabi Nu, Wilson ME et al. – Bilateral Idiopathic Brown's Syndrome With Delayed Onset in the Second Eye. J AAPOS 2000(3); 4: 158-63.
(8) Yarangumeli A CI, Kural G – Brown's Syndrome With Cyclic Characteristic: Case Report and Review of Physiopathologic Mechanism. J Pediatr Ophthalmol Strabismus 1995; 32: 243-47.

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