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BROWN'S SYNDROME, CURRENT CONCEPTS AND A CLINICAL REVIEW OF TWENTY CASES

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Abstract

Twenty cases of Brown's syndrome (two of which were acquired) who presented to the Eye Clinic at The Children's Hospital during a 12 month period from 1987 to 1988 are reviewed. The findings, which are similar to other published studies of this condition, show that the syndrome is generally a benign entity with a comparitively low incidence of strabismus and amblyopia. However, the examiner must be alert to the possibility of associated general and ocular conditions.

Key words: *Brown's syndrome, superior oblique tendon sheath syndrome, strabismus.*

The restrictive movement of Brown's syndrome (superior oblique tendon sheath syndrome) is thought to be due to an abnormality of the superior oblique tendon, its sheath or the Trochlea, preventing free passage of the tendon through the trochlea during inferior oblique action. Brown originally postulated in 1950[1] that the course was that the check ligament of the anterior tendon sheath of the superior oblique muscle was short and tight, possibly secondary to a congenital inferior oblique weakness. Electromyography studies, however, have shown that inferior oblique function is normal.[2]

Brown, in 1973 described two main groups: (i) true — which was congenital, permanent, and due to a congenital shortening of the anterior tendon sheath of the superior oblique, and (ii) simulated — where the anomaly could be permanent or intermittent, was acquired and had various aetiologies. Congenital cases which did not appear to be due to a shortened anterior tendon sheath were included.[3]

Parks and Brown describe an abnormal tendon, which is lacking the normal elasticity, to better explain a variety of clinical findings.[4]

Abnormal insertions of the superior oblique have been described in congenital cases where presentation was late in childhood, suggesting progression of the anomaly. Sandford-Smith postulates an abnormal relationship between tendon sheath and trochlea, secondary to an abnormal insertion, causing general "wear and tear" and secondary tendon swelling.[5]

It is also thought that adhesions between the sheath and tendon in its anterior parts, or, swelling or a nodule in the tendon behind the trochlea, is the mechanism leading to the acquired forms of Brown's syndrome.[6] This would allow movement of the tendon through the trochlea when the muscle is actively contracting but would prevent movement in the opposite direction.

Acquired Brown's syndrome is usually due to inflammation or trauma. Inflammatory changes may be due to local inflammation in the orbit; or part of more generalised inflammatory diseases such as rheumatoid arthritis and tenosynovitis.[5] Sinusitis and diseases of the nasopharynx can lead to a Brown's syndrome secondary to fibrous proliferation about the

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trochlea, the tendon and its sheath preventing free movement.[7,8]

Wright reported a clinicopathologic study of a patient with acquired inflammatory Brown's syndrome.[9] The superior oblique tendon, trochlea, and anterior superior oblique muscle were surgically-removed and studied by light microscopy. The entire specimen was normal, without signs of inflammation or intrasheath scarring. The only abnormal finding was perisheath adhesions anterior to the trochlea. This indicated that the cause of acquired inflammatory Brown's Syndrome may, in many cases, be due to scarring around the tendon sheath, rather than an intrasheath pathologic condition.

The acquired form of the syndrome can result from direct trauma to the region of the trochlea. A similar appearance is seen after some orbital floor fractures, frontal ethmoidal fractures, and crush fracture of nasal bone.[10,11] Trimble, Kelly and Mitchell have reported two cases which followed windscreen glass injuries, which initially presented as superior oblique weakness, but at about one month after injury the disorder spontaneously changed to a typical Brown's syndrome.[12]

Brown's syndrome has also followed frontal sinus surgery due to secondary fibrous proliferation about the trochlea and superior oblique tendon, and has been reported after superior oblique tucking.[11,13]

Booth-Mason and Kyle report the presentation of a 62 year old man with Brown's syndrome due to an orbital metastatic deposit.[14] There was a four month history of diplopia in elevation, and bi-frontal headaches. There was no relevant past medical history — although he had been a heavy smoker. There was no history of ocular disease; nor anything else found on clinical examination. General medical and neurological examinations were normal. Routine haematological, biochemical and immunological tests were negative. Skull and orbital X-rays were normal, but a chest X-ray showed an opacity, which on biopsy was found to be an undifferentiated carcinoma. A computerised tomographic scan showed a mass in the region of the superior oblique, with no bony erosion around the mass. This strongly suggested a metastatic deposit. Radiotherapy to the orbit was tried, but its effects on motility could not be accurately assessed because the patient deteriorated rapidly due to cerebral metastases, and died shortly afterwards.

Both congenital and acquired Brown's syndrome may be intermittent when the "click" phenomenon may be present.[5] An audible or palpebral snap may be noted by the patient or examiner during attempted upgaze. Following sustained effort or pressure over the trochlea, full elevation is possible. The "click" phenomenon is regarded as a stage in the resolution of the condition, Waddell postulating that the mechanisms being either enlargement of the trochlea with growth, or "wearing down" of the swelling with time.[15] In no cases in the following study from The Children's Hospital, could a "click" be recognised.

Pittke relates Brown's syndrome to a pseudoparesis of the inferior oblique muscle and terms it the proximal click syndrome due to abnormal tendon, anterior to the trochlea.[16] He has described a case of distal click syndrome which appears as a pseudoparesis of the superior oblique muscle, where the tendon is affected beyond the trochlea. A 25 year old woman presented for evaluation of "haziness of vision" in her right eye. There was no past ocular history, but fundoscopy revealed scarring and irregularity of the right optic disc. The right visual field showed an inferior temporal "horn-like" scotoma probably due to an old chorioretinitic lesion. There was a pseudoparesis of the right superior oblique muscle, which disappeared by blinking forcibly or by slight shaking of the head. When movement of the right eye stopped, the patient reported brief onset of diffuse, blurred vision, but without diplopia. The patient was unaware of the mechanism by which she compensated for the impaired superior oblique.

With regard to inheritance, Moore, Walker and Taylor[17] report bilateral Brown's syndrome in two siblings from their own experience, and report on six other apparent familial cases with more than one member of a family being affected with Brown's syndrome. In all the reported families the numbers of affected

TABLE I
Comparative incidence (in %) of features associated with Brown's Syndrome (% rounded to nearest whole figure)

This study	Male/Female	Bilateral	Unilateral (R, L)	Forced Primary Gaze			CHP, not binocular	No CHP not binocular
				Binocular	Hypotropia	Intermittent exotropia/ hypotropia		
This study (20 cases)	65/35	20	80 (60/40)	50	15	5	15	15
Clarke and Noel[22] (28 cases)	57/43	11	89 (36/64)	46	18	7	29	—

individuals are small, therefore, speculation on the mode of inheritance is difficult. It has been suggested that an embryologic insult occurs early; the finding of Brown's syndrome in more than one member suggesting that this early insult may have a genetic component.

Eleven cases of inferior oblique palsy were presented by Pollard and the benign nature of this entity stressed.[18] The inferior oblique is the least likely of all the extraocular muscles to be involved in an isolated paresis. Burian and von Noorden[19] have reported that it was the rarest extraocular muscle to be paralysed, and this has been reiterated by von Noorden and Oliver.[20] Of Pollard's cases the aetiology was attributed to congenital, traumatic, or presumed vascular nature. No cases of central nervous system tumour, infection, myasthenia gravis or diabetes were diagnosed. Marlow in 1923 reported one case occurring after a sinus infection, another by a small tumour of the orbit, and another with central nervous system infection due to syphilis.[21] There is little literature reporting inferior oblique muscle paresis.

Although no pathology was found in Pollard's study, patients who present with such must have a full work up — for example, a small stroke that causes inferior oblique paresis might be the harbinger of further vascular disease.

Twenty cases of Brown's syndrome presented at the Eye Clinic at The Children's Hospital over a 12 month period from 1987-1988. Of these 65% were female, and 35% male. Eighty percent were unilateral, of which 40% involved the left eye, and 20% were bilateral. This compares with the similar study by Clarke and Noel[22] who reviewed 28 cases of Brown's syndrome and reported a 57% male predominance, with only 10.7% being bilateral. Of the unilateral cases the left eye was affected in 54%.

In this series, difficulty in elevation was the presenting problem in 35% of cases. Reported horizontal strabismus was the next most frequent cause for concern in 15%, although all of these cases were actually heterophoric. Other presenting problems included compensatory head posture (CHP), school medical service referral, eye injury and a family history of eye problems (although not Brown's syndrome).

Eighteen (90%) of the cases were congenital. Of the two acquired cases, one was due to inflammation from juvenile rheumatoid arthritis which responded well to steroids. The second followed a kick to the face, although no associated orbital fracture was found.

Some binocularity could be demonstrated in 13 (65%) cases, and 10 of these had a CHP (neither of the acquired cases developed a CHP). 50% were binocular in forced primary gaze, which is similar to the Clarke and Noel study. Other similar findings in the two studies are detailed in Table 1. In addition, in this series 20% had esotropia with hypotropia in forced primary gaze, and 10% had constant esotropia.

Congenital Brown's syndrome is usually a benign entity. Amblyopia occurred in six (30%) of these cases, of which two could be classified as strabismic amblyopia and the other four ametropic amblyopia, who required correction of small to moderate amounts of hypermetropia and astigmatism. Two cases required surgery for horizontal strabismus, and one to lessen a CHP. This patient interestingly, had good binocular vision both with and without the CHP.

Congenital cardiac anomalies have been reported in association with this syndrome.[23] Conditions associated with the acquired form include rheumatoid arthritis[3] juvenile rheumatoid arthritis,[24] sinusitis and diseases of the nasopharynx,[11] tenosynovitis,[5] metastatic deposits[14] and trauma.[11]

Associated problems occurring in this series included developmental delay, microcephaly, mild dysmorphia, mild pulmonary stenosis (in the congenital cases) and juvenile rheumatoid arthritis and trauma (in the acquired cases).

Very few ocular anomalies are associated with this syndrome, but ptosis and hypertelorism have been reported.[23] Of the 20 cases in this series, one had lid coloboma and another had asymmetric orbits and lids.

CONCLUSIONS

This series shows similarities with other published studies on Brown's syndrome. It can be seen that it is generally a benign entity, but one must be always alert to the possibility of sinister signs which may also occur.

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References

1. Brown HW. Congenital structural muscle anomalies. In: Allen JH, ed. Strabismus Ophthalmic Symposium. St Louis: CV Mosby, 1950; 205-236.
2. Catford VG, Dean Hart JC. Superior oblique tendon sheath syndrome: an electromyographical study. Br J Ophthalmol 1971; 55: 155-160.
3. Brown HW. True and simulated superior oblique tendon sheath syndromes. Doc Ophthalmol 1973; 34: 123-126.
4. Parks MM, Brown M. Superior oblique tendon sheath syndrome of Brown. Am J Ophthalmol 1975; 1: 82-86
5. Sandford-Smith JH. Superior oblique tendon sheath syndrome and its relationship to stenosing tenosynovitis. Br J Ophthalmol 1973; 57: 859-864.
6. Roper-Hall MJ, Roper-Hall G. The superior oblique "click" syndrome. Orthoptics, ICS 245. Excerpta Medica, Amsterdam. 360-366.
7. Brown HW. Strabismus in the adult. Haik GM (ed). Strabismus Symposium of the New Orleans Academy of Ophthalmology. St Louis: CV Mosby, 1962: 248-273.
8. Clarke E. A case of apparent intermittent overaction of the left superior oblique. Br Orthopt J 1966; 23: 116-117.
9. Wright KW, Silverstein D, Marrone AC, Smith RE. Acquired inflammatory superior oblique tendon sheath syndrome. A clinicopathologic study. Arch Ophthalmol 1982 (Nov); 100: 1752-1754.
10. Zipf RF, Trokel SL. Simulated superior oblique tendon sheath syndrome following orbital floor fracture. Am J Ophthalmol 1973; 75: 700-705.
11. Blanchard CL, Young LA. Acquired inflammatory superior oblique tendon sheath (Brown's) syndrome. Report of a case following frontal sinus surgery. Arch Otolaryngol 1984; 110: 120-122.
12. Trimble RB, Kelly V, Mitchell M. Acquired Brown's syndrome. In: Ravault AP, Lenk M (eds). Trans v Int Orthoptic Congress Lyons: LIPS, 1983: 267-273.
13. Expression Y, Hudelo J, Weiss JB. Syndrome et pseudo-syndrome de Brown. Bull Soc Ophthalmol Fr 1969; 69: 812-815.
14. Booth-Mason S, Kyle GM, Rossor M, Bradbury P. Acquired Brown's syndrome: an unusual cause. Br J Ophthalmol 1985; 69: 791-794.
15. Waddell E. Brown's syndrome revisited. Br Orthopt J 1982; 39: 17-21.
16. Pittke EC. The proximal and distal click syndrome of the superior oblique tendon. Graefe's Arch Clin Exp Ophthalmol 1987; 225: 28-32.
17. Moore AT, Walker J, Taylor D. Familial Brown's Syndrome. J Ped Ophthalmol Strab 1988; 4(25): 202-204.
18. Pollard ZF. Inferior oblique paresis: a benign entity. Ann Ophthalmol 1986; 18: 17-180.
19. Burian H, von Noorden G. Paralytic strabismus. In: Burian H, von Noorden G. Binocular Vision and Ocular Motility. St Louis: CV Mosby, 1974: 357.
20. Oliver P, von Noorden G. Results of superior oblique tenectomy in inferior oblique paresis. Arch Ophthalmol 1982; 100: 581-584.
21. Marlow SB. Isolated paralysis of the inferior oblique. Arch Ophthalmol 1923; 52: 12-24.
22. Clarke WN, Noel LP. Brown's Syndrome: fusion status and amblyopia. Can J Ophthalmol 1983; 18(APR): 118-123.
23. Rao VA, Sood GG and Santoshkumar. Brown's syndrome with congenital ptosis. Ind J Ophthalmol 1982; 30: 57-59.
24. Searle CM, Horne SM. Acquired Brown's syndrome: a case report. Aust Orthopt J 1984; 21: 43-46.
25. Jackson OB (Jnr), Nankin SJ, and Scott WE. Traumatic simulated Brown's syndrome: a case report. J Ped Ophthalmol Strab 1979; 16: 160-162.