Oman Medical Journal (2013) Vol. 28, No. 4
DOI 10. 5001/omj.2013.84

A Case of Ocular Torticollis

Doddaiah Narayanappa, Hassan Srinivasamurthy Rajani, Timmanahalli Girish Anita, and Nagaraj Rashmi

Received: 12 Feb 2013 / Accepted: 03 Apr 2013

Abstract

A 1-year-and-2-month-old male baby was brought to Pediatric out patient department with head tilt to left side, chin rotated to right side with full range of neck movement, right eye hypertropia increased on left gaze, and not associated with any other musculoskeletal abnormalities. Diagnosis of ocular torticollis secondary to right superior oblique muscle palsy was made.

Keywords: Ocular; Superior oblique muscle; Torticollis.

Introduction

The term 'torticollis' means twisted neck and is derived from the Latin words 'tortus' and 'collum'.[1] Torticollis is a clinical symptom and a sign characterized by a lateral head tilt and chin rotation towards the side opposite to the tilt. The reported incidence of congenital torticollis is 0.3% to 2%.[2] Congenital muscular torticollis associated with contracture of the sternocleidomastoid muscle (fibromatosis colli or sternocleidomastoid tumour),[3] is the most common etiology of torticollis in infants.[4] Unusual causes are eye muscle weakness, Sandifer's syndrome resulting from gastroesophageal reflux, neural axis abnormalities, and benign paroxysmal torticollis. We are hereby presenting a case of torticollis initially misdiagnosed as muscular in origin but subsequently found to be secondary to ocular muscle palsy.

Case Report

A 1-year-and-2-month-old male baby, first child of a non-consanguineously married couple with uneventful antenatal, natal (no history of difficult delivery, birth trauma), and postnatal a history with normal developmental milestones was brought with history of tilting of the neck to the left side noticed by parents since the age of 4 months. There was no history of head injury, no history of feeding difficulties, and no family history of any ocular disorders. Torticollis secondary to contracture of the sternocleidomastoid muscle was diagnosed elsewhere and physiotherapy was instituted with no improvement.

Doddaiah Narayanappa ✉ **, Hassan Srinivasamurthy Rajani, Nagaraj Rashmi**
Department of Pediatrics, JSS Medical College, JSS University, India.
E-mail: sinchabhi@yahoo.com

Timmanahalli Girish Anita
Department of Ophthalmology, JSS Medical College, JSS University, India.

On examination, head tilt to the left side with chin rotated to the right side was present with full range of neck movement assessed by active head turning during visual tracking.[5] On inspection of the neck, there was no asymmetry of skin folds. All superficial cervical muscles were palpated with no evidence of mass or contracture of the sternocleidomastoid muscle. No facial asymmetry or plagiocephaly noted. Musculoskeletal examination was unremarkable (no CTEV, congenital dislocation of hip, scoliosis). Orthopedic opinion was taken. X-ray of the cervical spine was normal.

On ophthalmological evaluation, right eye hypertropia was present. In forced primary position (when his head was held straight), there was right eye hypertropia. In right gaze, there was no change in the hypertropia, but in left gaze, increase in hypertropia was noted suggestive of right superior oblique muscle palsy. This is a positive Bielschowsky head tilt test and is indicative of paralysis of the superior oblique muscle. Diagnosis of ocular torticollis secondary to right superior oblique muscle palsy was made and the patient was referred to an ophthalmologist for further management.

Discussion

The congenital and developmental causes of torticollis in children have been classified as osseous, nonosseous or neurogenic.[4,6] The prevalence of nonmuscular causes of torticollis in children could be as high as 18%.[7] The common ocular causes of torticollis in infancy are congenital paralytic squint and congenital nystagmus. Paresis of the superior oblique muscle results in head tilt away from the side of impairment and is the most common cause of ocular torticollis.[1,8] Other ocular causes are lateral rectus muscle palsy and Duane's syndrome (a congenital condition simulating lateral rectus paresis).

The superior oblique muscle is a fusiform muscle originating in the upper, medial side of the orbit, innervated by the trochlear nerve. Its primary action is intorsion (internal rotation), the secondary action is depression (primarily in the adducted position), and the tertiary action is abduction (lateral rotation). Patients with a paralytic squint adopt an abnormal head posture to maintain binocular vision and avoid diplopia. The exact mechanism depends on the affected muscle; the head may be placed so that the eyes are directed away from its field of action, thereby allowing ocular alignment to be maintained. Patients with paresis of a horizontally-acting extra-ocular muscle may achieve this with face

turn alone, but a combination of face turn and chin elevation or depression may be necessary to correct for an abnormal vertically-acting muscle, and head tilt may also be found.

The Parks-Bielschowsky Three Step Test attempts to determine the paretic muscle by performing alternate cover testing in different head positions. This test only works in cases of a single paretic muscle. Since the superior oblique is the vertical muscle most commonly affected, this is basically a test for dysfunction of the superior oblique. The first step is to determine which eye is hypertropic. In this child, the right eye was hypertropic. The second step is to determine whether the hypertropia is greater in left or right gaze. Hypertropia due to superior oblique paralysis is greater on gaze to the contralateral side. In this child, hypertropia increased on left gaze indicating right superior oblique paresis. The third step is to determine whether the hypertropia is greater in left or right head tilt. Hypertropia due to superior oblique paralysis is greater in a head tilt to the ipsilateral side.

Some features other than ocular pathology may help to distinguish ocular from non-ocular torticollis.[1] Fibromatosis colli, the most common etiology of torticollis in infants,[4] is associated with history of birth trauma, difficult delivery (especially use of forceps), or breech delivery,[3] which were not present in our case. Usually, ocular torticollis presents later than non-ocular, after head control and binocular vision are established. In this case as well as in few other studies,[1] abnormal head posture was noted at an early age. Thus, age at onset may not be a useful distinguishing feature. Family history of nystagmus in a child with torticollis may be helpful, but as ocular abnormalities are inherited in a complex multifactorial fashion, a family history of these conditions may not be predictive. The full range of neck movement in ocular and restriction in the non-ocular group may be helpful,[3] but longstanding torticollis of ocular origin may eventually produce secondary changes in the neck muscles and restrict movement. Occlusion of one eye may help to differentiate some patients with ocular cause, but in longstanding torticollis with secondary neck changes, or when an ocular torticollis is for reasons other than maintaining binocular vision, the test has limited value.

Conclusion

Hence, a high index of suspicion is necessary for early diagnosis of ocular torticollis. In late cases, it may persist even after treatment of ocular defect, and even secondary plagiocephaly has been reported.[1] As it is not easily possible to clinically distinguish ocular from non-ocular causes, we recommend that all patients with torticollis and no clear orthopedic cause must be referred for ocular assessment.

Acknowledgements

The authors reported no conflict of interest and no funding was received for this work.

References

1. Williams CR, O'Flynn E, Clarke NM, Morris RJ. Torticollis secondary to ocular pathology. J Bone Joint Surg Br 1996 Jul;78(4):620-624.
2. Herman MJ. Torticollis in infants and children: common and unusual causes. Instr Course Lect 2006;55:647-653.
3. Khalid S, Zaheer S, Wahab S, Siddiqui MA, Redhu N, Yusuf F. Fibromatosis Colli: A Case Report. Oman Med J 2012 Nov;27(6).
4. Berlin H. The differential diagnosis and management of torticollis in children. Phys Med Rehabil Clin N Am 2000;14:197-206.
5. Persing J, James H, Swanson J, Kattwinkel J; American Academy of Pediatrics Committee on Practice and Ambulatory Medicine, Section on Plastic Surgery and Section on Neurological Surgery. Prevention and management of positional skull deformities in infants. Pediatrics 2003 Jul;112(1 Pt 1):199-202.
6. Loder RT. The cervical spine. In: Morrissy RT, Weinstein SL, eds. *Lovell and Winter's Pediatric Orthopedics*, 5th ed. Philadelphia: Lippincott Williams & Wilkins; 2001:799–836.
7. Ballock RT, Song KM. The prevalence of nonmuscular causes of torticollis in children. J Pediatr Orthop 1996 Jul-Aug;16(4):500-504.
8. Cheng JC, Wong MW, Tang SP, Chen TM. MPhil, Shum SLF, Wong EMC. Clinical determinants of the outcome of manual stretching in the treatment of congenital muscular torticollis in infants. Bone Joint Surg. 2001;83:679-687.