Torticollis, head bobbing and oscillatory eye movements in a 14 year old child

Sir,

Spasmus nutans is a rare condition, characterized by rapid nystagmus in the horizontal plane, which may be disjunctive or monocular, head bobbing, and torticollis. It usually starts around 6 months of age and resolves spontaneously by 3-4 years. In general, the condition is self-limiting and the prognosis is good. Occasionally, it is consequent upon structural brain lesions or diseases of the anterior visual pathway, achromatopsia, or retinal dystrophy. We report one such case and present the video as well.

A 14-year-old boy presented to our outpatient department with involuntary abnormal movements of the eyeballs and deviation of the neck to the right side almost since birth. These features were observed by his parents and the subject was unaware of them, though he mentioned that he had some difficulty in seeing distant objects. He was born of nonconsanguineous marriage; his birth, developmental and intellectual milestones were uneventful; and the illness was nonprogressive. On examination, his higher mental functions were within normal limits. There was minimal spontaneous nystagmus on forward gaze which was aggravated in both lateral gaze, right being more than the left, along with a torsional component. Fundoscopic examination was within normal limits and the pupils were normal in size and smartly reacting to light. The deep tendon reflexes were within normal limits and the plantar response was flexor. There was no evidence of any cerebellar dysfunction or any other involuntary movement. Somatic sensory and posterior column functions were within normal limits.

It was observed that the patient had head tilt to the right side along with slight prominence of the left-sided sternocleidomastoid muscle. Very minimal nodding of the head in the anteroposterior direction was seen, which was better felt when the hand was placed over his head. The patient's features were videoed for future documentation [Video].

Imaging study of brain and craniovertebral junction, and the visual evoked potential studies were within normal limits. Considering the classic triad of nystagmus, head bobbing, and torticollis in a young subject, the diagnosis of 'spasmus nutans' was entertained. Since there is no specific management for this condition and the condition is benign in nature, the patient was reassured.

The nystagmus in spasmus nutans is intermittent and consists of fine, pendular, and predominantly horizontal oscillations of high frequency up to 15 Hz.[1-3] The head nodding is also intermittent and it is believed that head nodding and torticollis are compensatory to nystagmus in order to improve vision. Additionally, the patients may shake their head vigorously in order to initiate the vestibule-ocular reflex for suppressing the ocular oscillations. Some patients may have an additional esotropia, which is purposeful to suppress the nystagmus, since this results in the head turning in the direction of the fixating adducted eye, the so called nystagmus blockade syndrome.[4] Usually, no other neurological deficit is observed in these subjects. However, various intracranial lesions may be manifested in some cases, such as cerebellar hypoplasia, third ventricular tumor, retinal dystrophy, and achromatopsia.[5,6] In about 2% cases, structural lesions affecting the anterior visual pathway, like, optic nerve glioma, optic atrophy, and relative afferent pupillary defect are the additional findings.[7]

Some investigators have observed concomitant deficiency of vitamin D and iron in some cases of spasmus nutans and these patients belonged to lower socioeconomic background.[8] Though the disorder resolves spontaneously, subtle abnormalities may persist in eye movement up to 12 years of age.[6] It is occasionally familial and have been reported in monozygotic twins as well.[9,10] Maybodi in his classic paper has suggested modulation of sympathomimetic modulation for infantile nystagmus.[11]

The pathogenesis of spasmus nutans has not been clearly elucidated. It is mainly idiopathic and is not accompanied by other neurological or extra-neurological abnormalities; although strabismus, developmental delay, cerebellar vermis hypoplasia, and congenital ocular motor apraxia of Cogan may be the associated conditions.[1,4]

The case in point was 14 years of age and no case to the best of our knowledge has been reported in the literature so far, who presented at such an old age.

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References

1. Norton EW, Cogan DG. Spasmus nutans: A clinical study of 20 cases followed for two years or more since onset. AMA Arch Ophthalmol 1954;52:442-6.
2. Gresty MA, Leech J, Sanders M, Eggars HA. Study of head and eye movement in spasmus nutans. Br J Ophthalmol 1976;60:652-5.
3. Weissman BM, Dell'Osso LF, Abel LA, Leigh RJ. Spasmus nutans: A quantitative prospective study. Arch Ophthalmol 1987;105:525-8.
4. Bradley's Neurology in Clinical Practice. In: Daroff RB, Fenichel GM, Jankovic J, Mazziotta JC, editors. 6th edn; 2012. p. 603.
5. Lavery MA, O'Neill JF, Chu FC, Martyn LJ. Acquired nystagmus in early childhood: A presenting sign of intracranial tumor. Ophthalmology 1984;91:425-53.

6. Kim JS, Park SH, Lee KW. Spasmus nutans and congenital ocular motor apraxia with cerebellar vermian hypoplasia. Arch Neurol 2003;60:1621-4.
7. Kiblinger GD, Wallace BS, Hines M, Siatkowski RM. Spasmus nutans-like nystagmus is often associated with underlying ocular, intracranial, or systemic abnormalities. J Neuropthalmol 2007;27:118-22.
8. Dail AI, Saygili O. Risk factors in spasmus nutans. Adv Clin Exp Med 2011;20:183-6.
9. Hoyt CS, Aicardi E. Acquired monocular nystagmus in monozygous twins. J Pediatr Ophthalmol Strabismus 1979;16:115-8.
10. Vassella F, Lutschg J, Mumenthaler M. Cogan's congenital ocular motor apraxia in two successive generations. Dev Med Child Neurol 1972;14:788-96.
11. Maybodi M. Infantile onset nystagmus. Curr Opin Ophthalmol 2003;14:276-85.

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