See-saw Nystagmus in Chiari Malformation Type I

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ABSTRACT

Purpose: To report a case of 25 year-old woman with see-saw nystagmus as a manifestation of Chiari malformation type I. Methods: A 25 year-old woman was referred with involuntary movements of both eyes. Assessments were for visual acuity and anterior and posterior segments of the eyes, and magnetic resonance imaging of head and brainstem. Result: See-saw nystagmus was evident. Best corrected visual acuity were 6/6 in both eyes. Anterior and posterior segments of the eye were normal. The diagnosis of Chiari malformation type I was confirmed by magnetic resonance imaging. Conclusion: See-saw nystagmus can be found in Chiari malformation type I. No specific treatment was given.

Keywords: Chiari malformation, see-saw nystagmus

INTRODUCTION

Chiari malformation type I (CMI) is a disorder characterized by hindbrain overcrowding into an underdeveloped posterior cranial fossa (PCF), it was first reported in 1890 by H. Chiari, a professor of pathology from Germany. This malformation was divided into three types according to the level of herniation: Type I, the mildest, to type III, the most severe.1-3

Prevalence of Chiari malformation (congenital or acquired) is 1 per 1000 to 1 per 5000. Chiari malformation type I is common, less severe, and treatable, most are asymptomatic. Incidence of asymptomatic Chiari are unknown.4

The radiographic criteria for diagnosing a congenital Chiari malformation type I is a downward herniation of the cerebellar tonsils greater than 5 mm below the foramen magnum.5

Chiari malformation type I (congenital or acquired) can be asymptomatic or neurologic signs and symptoms may present commonly associated with direct neural compression at the craniovertebral junction or with cerebrospinal fluid flow disturbances. Although presentation typically occurs in middle age, it may start in childhood or infancy. It may cause sudden infantile death syndrome. Conversely, Chiari malformation type I often can be asymptomatic and tonsillar descent is often regarded as an incidental neuroradiological finding.4

One of the ocular disturbance found in Chiari malformation type I is nystagmus. See-saw nystagmus is a form of disconjugate nystagmus in which one eye elevates and intorts while the other eye depresses and extorts, resembling a see-saw movement.1-3 Nystagmus is generally difficult to treat, although successful forms of therapy have been reported for some forms.7

CASE REPORT

A 25 year-old woman was referred to ophthalmology department with involuntary movement on both eyes since 3 years.
LAPORAN KASUS

No history of ophthalmic disturbance. On examination, visual acuity on right and left eye were 6/15 and 6/12 respectively. Perception of light was normal in both eyes. Fundi examination using indirect and fundus photograph revealed normal imaging in both eyes. No history of significant neurological findings, no history of headache but there’s a history of amenorrhea for about 3 months. The examination of eye movement was normal, but see-saw nystagmus was evident. Patient underwent neuroimaging and revealed a herniation of cerebellar tonsil.

The patient was diagnosed with Chiari malformation type I. The patient was given spectacles with spheric negative lenses (S-0.75) on both eyes which improved her visual acuity to 6/6 on both eyes. No other treatment was given.

DISCUSSION
Chiari malformation type I (CMI) is a disorder of uncertain origin that has been traditionally defined as downward herniation of the cerebellar tonsils through the foramen magnum. In contrast to other Chiari malformations, CMI tends to present in the second or third decade of life and it is sometimes referred to as the “adult-type” Chiari malformation.7 Clinical symptoms of this malformation consists of symptoms of spinal cord dysfunction due syringohydromyelia (64%), including scoliosis, dissociation of pain and temperature sensory sensation, dysesthesia in extremities, limb rigidity, urinary incontinence, and motor weakness; symptoms of brainstem disorder (28%) such as sore neck/head, hoarseness, palate dysfunction, tongue ataxia, nystagmus. Rare acute neurologic symptoms may present.5,6 The patient was asymptomatic until confirmed by neurologist. A study on 364 symptomatic CMI patient showed that ocular disturbances were reported by 283 patients (78%) (Table).1

In this case, we only found see-saw nystagmus as a clinical manifestation since the last three years without jumping image (oscillopsia) as regularly found in acquired nystagmus. This is probably because the patient has had the nystagmus since childhood and already been adapted or may not be disturbed because it only presents in extreme lateral gaze.

See-saw nystagmus has been seen with tumors of the parasellar region and diencephalon, brain-stem vascular lesions, syringobulbia, and after trauma, very rarely found in Chiari malformation type I, demonstrated in this patient. An association between see-saw nystagmus and Chiari malformation is potentially important because early diagnosis and decompression may improve neurologic function and prevent further deterioration.

Chiari malformation type I diagnosis can be established by magnetic resonance imaging (MRI). Therapy of choice is continued clinical monitoring and surgical intervention. Some cases have rare spontaneous resorption of syringomyelia. Clinically asymptomatic patient without syringomyelia should be observed. Symptoms progression or significant clinical disorder are an indication for surgery.8,10 Surgery procedures consist of posterior fossa craniectomy and cervical laminectomy for decompression.

CONCLUSION
See-saw nystagmus can be found as first presentation of Chiari malformation type I. Since our patient was clinically asymptomatic and syringomyelia was not found, no specific treatment was offered.
REFERENCES


