Case Report

Chiari type 1.5 malformation with the right upper limb monoparesis and Horner's syndrome: A case report

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ABSTRACT

Chiari 1.5 malformation is a rare condition characterized by caudal descent of the brain stem, together with the descent of the cerebellar tonsils. This is a case report of a 36-year-old female who had right upper limb monoparesis and Horner's syndrome since birth and presented with complaints of vertigo and unsteady gait. On examination, revealed ataxia and nystagmus and magnetic resonance imaging showed Chiari type I.5 malformation with associated mild communicating hydrocephalus, syringobulbia, and syringomyelia.

Key words: Chiari malformation, Downbeat nystagmus, Horner's syndrome, Hydrocephalus, Syringobulbia, Syringomyelia

Chiari malformation is a rare disease with prevalence rates of 0.1–0.5% and female predominance (1.3:1) [1]. Chiari malformation is characterized by herniation of cerebellar vermis for more than 3 mm below the foramen magnum. Such malformation can be either congenital or acquired through trauma [2]. There are six recognized Chiari types. Chiari types I, II, and III share the common characteristic of varying degrees of herniation of hindbrain derivatives out of the posterior cranial fossa, of which Chiari type I is the most common. Chiari IV malformation involves cerebellar hypoplasia or aplasia with no herniation of the hindbrain. Chiari Type 0 has recently been recognized as a form of Chiari where the herniation does not meet normal criteria but a syrinx is present and causes symptoms [3]. Similarly, Chiari 1.5 malformation is a newer terminology. It is also called a bulbar variant of Chiari I malformation. Tonsillar herniation >12 mm suggests Chiari 1.5 malformation (>6 mm suggests Chiari I malformation). It is considered a progression of Chiari I malformation [4]. It is thought to be less common than the Chiari I malformation; although, the exact incidence of Chiari 1.5 malformation is still unknown [5]. Here, we present a rare case report of a patient with Chiari 1.5 malformation.

CASE REPORT

A 36-year-old female patient was having right upper limb weakness and drooping of the right upper eyelid since birth for which, she has not consulted any doctor and was neglected as a birth injury. The weakness was non-progressive. It was not involving the left upper limb or lower limbs. There was no history of bowel and bladder involvement. The patient developed vertigo and unsteadiness of gait in the last 3 months, which was gradually progressive in nature. The patient also developed an occipital headache which became severe in the last week.

General examination was unremarkable and the patient was vitally stable. On examination, the patient had thinning of the right thenar and hypothenar muscles and clawing of fingers of the right hand (Fig. 1). Power of grade 3/5 was present in the right upper limb in all joints, with associated hypotonia. The power was grade 4/5 in the right lower limb and full power in the left upper limb and lower limb. The deep tendon reflexes were absent in the right upper limb and exaggerated in the right lower limb. The deep tendon reflexes were normal in the left side of the body. The sensory system was normal. Cranial nerve examination was normal. The patient also had an ataxic gait. Other cerebellar signs were absent. The right-sided eye had ptosis, enophthalmos, and miosis which were suggestive of Horner's syndrome (Fig. 1). Downbeat nystagmus was present in the right eye. Spine examination showed thoracic scoliosis.

Blood investigations were normal. Magnetic resonance imaging (MRI) brain and cervical spine were planned as the patient had downbeat nystagmus and ataxia and the patient had lower motor neuron signs in the right upper limb and upper motor neuron signs in the lower limb. MRI was suggestive of Chiari type I.5 malformation with peg-like caudal descent of tonsils into the upper cervical canal by 1.5 cm (Fig. 2). There

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was also associated descent of the brainstem with the lower part of the medulla lying below the foramen magnum. Mild upstream dilation of bilateral second, third, and fourth ventricles (mild communicating hydrocephalus) was present. Mild retropulsion of the odontoid process and relatively small posterior fossa was present. Syringobulbia was also present and syringomyelia was found to be extending from C3 to T1 with the associated asymmetric and atrophic spinal cord.

The patient was given symptomatic management and was referred to a neurosurgeon for further evaluation. The patient was lost to follow-up.

DISCUSSION

The etiology of Chiari 1.5 malformation is unknown. One of the theories proposed for the pathogenesis of the condition involves the presence of difference of pressure between cranial and spinal subarachnoid space which causes abnormal fetal brain development during pregnancy. There is thought to be some genetic predisposition to Chiari, since the defect can rarely be found running in families but no specific genetic cause has not been identified. In our patient's case, a family history of similar illness was absent. Some rare conditions that result in abnormal bone growth such as craniosynostosis, skeletal dysplasia, and achondroplasia can also present with similar symptoms and structural abnormalities [3].

Figure 1: Clinical features of the right upper limb monoparesis (claw hand and thenar and hypothenar muscle wasting) and Horner's syndrome (ptosis and enophthalmos)

The essential difference between Chiari I and 1.5 is the presence of caudal descent of the brainstem in the latter along with tonsillar ectopia [3]. These two clinical conditions share many similar features and no signs or symptoms can differentiate the two. Type 1 Chiari malformation typically presents in the second or third decade of life (25–30 years) [6]. However, younger age at presentation and more severe symptoms like bulbar signs are more common in Chiari 1.5. However, our patient presented at the age of 36 years. The patients in Chiari type I are asymptomatic in 30% of cases and headache is the most common symptom, but the visual symptoms are present in up to 80% of patients [7,8]. Our patient also had an occipital headache.

Other symptoms of syringobulbia included neck pain, cranial nerve palsy, limb weakness, or dysesthesia, Horner syndrome, ataxia, and respiratory disorders. Limb weakness, whether unilateral or bilateral, was also a common symptom. Diplopia and ptosis were also common. Syringomyelia is present in up to 80% of cases. Scoliosis commonly occurs in the setting of Chiari I malformation and even more frequently in the setting of Chiari I malformation with syringomyelia [9]. However, the exact incidence of scoliosis in Chiari 1.5 malformation is unavailable. Our patient also had thoracic scoliosis which was evident on clinical examination and in chest X-ray.

Accompanying syringobulbia is much rare (<5% of cases) [8]. Neurological examinations usually revealed tendon hyperreflexia, although hyporeflexia was also occasionally found. Ataxia, gait disturbance, nystagmus, and other cerebellar signs were also common. Hydrocephalus is uncommon and rare in Chiari malformation. Our patient had hydrocephalus also. RS Tubbs *et al*. conducted a study among 130 symptomatic pediatric Chiari I malformation patients and 22 of them were Chiari 1.5 malformation patients. Clinical manifestations and symptomatic outcomes were compared between the groups and were found to be similar in the two groups in terms of the degree of tonsillar herniation (Chiari I malformation vs. Chiari 1.5 malformation: 7 mm vs. 12.7 mm), the angulation of the odontoid process (98.0°

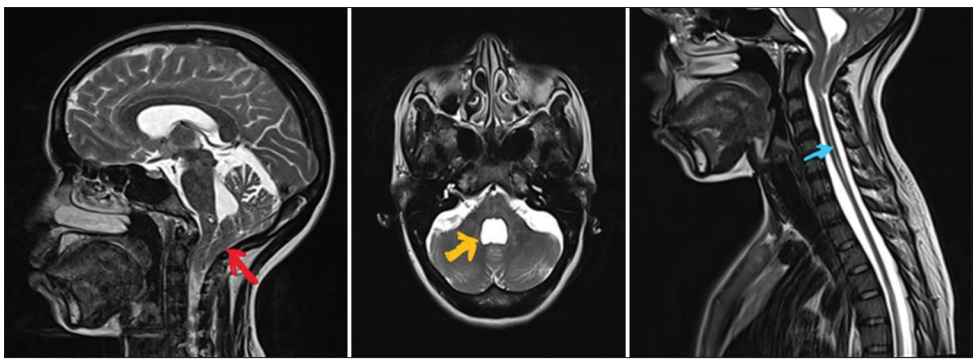

Figure 2: Magnetic resonance imaging findings of tonsillar herniation (red arrow), syringobulbia (yellow arrow) and syringomyelia from C3 to T1 (blue arrow)

vs. 84.4°), hydrocephalus (11% vs. 9.1%), syringomyelia (58% vs. 50%), scoliosis (17% vs. 27%), and post-operative improvement of symptoms (83% vs. 82%) [5].

MRI is the imaging modality of choice; sagittal image is the best for assessing the presence of Chiari 1.5 malformation. Axial images through the foramen magnum show crowding of the posterior fossa with cerebellar tonsil and brain stem. Neuroimaging feature of Chiari 1.5 is the descent of obex and cerebellar tonsils below the foramen magnum [3].

In the case of a severe advanced form of Chiari 1.5 malformation, sufficient posterior fossa decompression including tonsillectomy for normalizing cerebrospinal fluid circulation around the foramen magnum is necessary. But have a less consistent response to surgical treatment, with a higher incidence of persistent syringomyelia [10]. Sometimes, repeated surgeries are required for the same. The patient mentioned in this case report presented at a late stage, so her prognosis was not good. Surgical intervention at this stage carries the risk of postoperative complications and recurrent surgeries might be needed. The prognosis could have been better if she presented at an earlier age and surgical intervention would have prevented progression to Chiari 1.5 malformation.

CONCLUSION

This is a rare case report of Chiari 1.5 malformation with late presentation. A high index of suspicion should be kept in patients with clinical features of cervical involvement, Horner's syndrome, and cerebellar signs for which, early surgical intervention should be given to them. This is an attempt to present a relatively rare and newer entity in the Chiari spectrum, analyze its manifestations, and create awareness among the fraternity to broaden the scope of research in this subject, which in our opinion is very important due to the differences in its management and prognosis of this condition.

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