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Case Report

Chiari zero malformation with syringobulbia ☆

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ABSTRACT

Chiari zero malformation is a relatively new and rare subtype of Chiari malformations. Most of the patients present with signs and symptoms of Chiari malformation without actual cerebellar tissue herniation, with or without syringomyelia. Furthermore, Chiari zero cases can be associated with syringobulbia in rare instances. We present a case of a 39-year-old patient diagnosed with Chiari zero associated with syringomyelia and syringobulbia.

Introduction

Chiari malformation (CM) is a collection of congenital malformations discovered by Hans Chiari in 1891 that generally affects children and young adults [1]. CM has an ambiguous pathophysiology. However, patients present with symptoms related to the herniation of cerebellar tissue through the foramen magnum, such as pain, weakness, atrophy, sensory deficits, and others [1,2]. Hans originally classified CM into 4 types (CM 1-4) with type one being the most encountered. However, 2 additional types were later described, type 0 and type 1.5 [3].

CM 1 is defined when the cerebellar tonsils herniate below the foramen magnum (>5 mm) into the cervical spinal canal but without the involvement of the brain stem [1,4]. In contrast, CM 2 involves the herniation of the medulla and fourth ventricle in addition to the cerebellar vermis [2]. Both of these variants are associated with myelomeningocele, which led to the development of a new variant, in which the tonsils and brainstem herniates without the presence of myelomeningocele, and it is called CM 1.5 [2,3]. Furthermore, CM 3 occurs when there is posterior encephalocele into which parts of the brain stem and cerebellum herniates and CM 4 is associated with hypoplasia of the tentorium along with hypoplastic or aplastic cerebellum [3].

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Chiari zero malformation (CM0), a rare Chiari subgroup described as an absence of hindbrain herniation, with the presence of syringomyelia that will eventually resolve after posterior fossa decompression [3]. However, syringomyelia was deemed not mandatory anymore in the diagnosis, as reported recently [4,5]. Rarely, CM0 may be associated with syringobulbia [5].

Herein, we present a case of a 39-year-old patient who has been diagnosed with Chiari zero malformation associated with syringomyelia and syringobulbia.

Case presentation

A 39-year-old patient presented complaining of progressive bilateral hand numbness and tingling of 2 years duration. He was previously diagnosed with obstructive sleep apnea and scoliosis. Lately, he starts complaining of left upper limb weakness and pain which were debilitating. He was vitally stable with a body mass index of 39.2 kg/m^2. On physical examination, there was no nystagmus, personality changes, enlarged tongue, or hiccups. In addition, it was not associated with neurological features or signs of increased intracranial pressure such as dysarthria, vomiting, abnormal muscle movement, hoarseness, dysphagia, or vision or hearing problems. There was a decrease in the left upper extremity power and pain sensation with numbness and tingling, without muscle twitching. The right upper extremity power was intact, along with the bilateral lower extremities. Moreover, the sensation was intact elsewhere. Magnetic resonance imaging (MRI) was ordered. It showed a low laying medullary obex without tonsillar herniation or hydrocephalous (Fig. 1). In addition, a large syrinx involving the whole cervical cord, with an extension of the syrinx to the medulla oblongata (syringobulbia) which measured 6 × 7 × 18 mm anteroposterior, transverse, and cephalocaudal respectively (Fig. 2), with no evidence of enhancing obstructive masses (Fig. 3). Moreover, the extension of the large syrinx involves all the dorsal cord, only the conus medullaris is

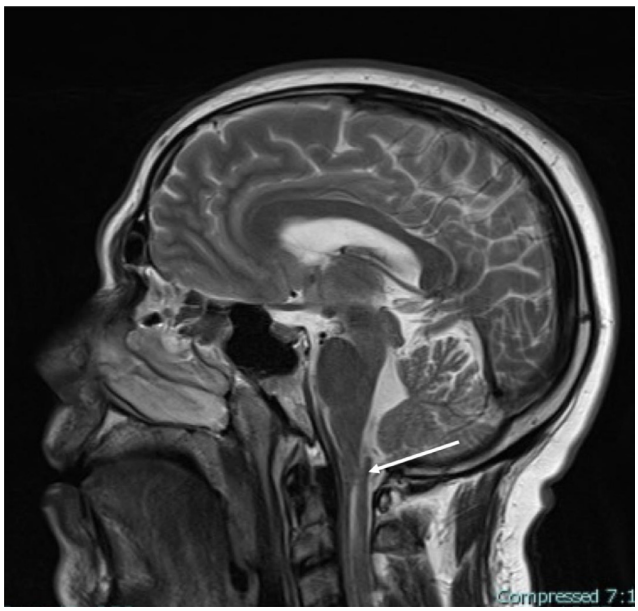

Fig. 1 – A sagittal T2 MRI of the brain showing a low lying medullary obex. No tonsillar herniation or Hydrocephalus.

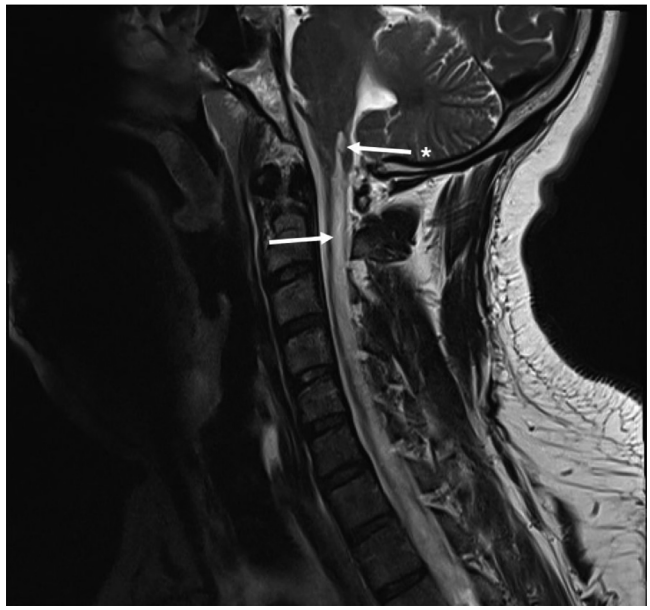

Fig. 2 – A sagittal T2 MRI of the cervical spine showing a large syrinx involving the whole cervical cord with an extension of the syrinx to the medulla oblongata (Syringobulbia) (star).

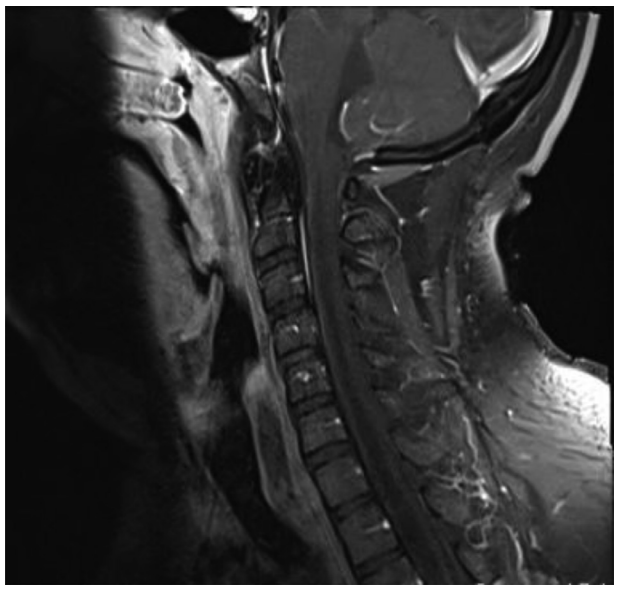

Fig. 3 – A contrast-enhanced T1 fat-sat MRI of the cervical spine showing no evidence of enhancing obstructive masses.

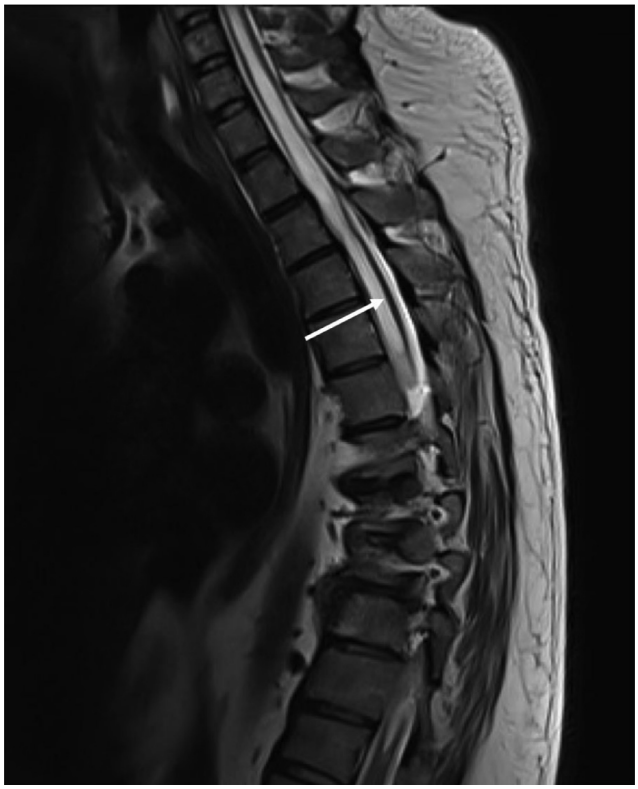

Fig. 4 – A sagittal T2 MRI of the dorsal spine showing an extension of the large syrinx to involve all the dorsal cord. Only conus medullaris is preserved.

preserved (Fig. 4). Depending on the aforementioned findings, the patient was diagnosed with Chiari zero malformation and referred to neurosurgery for further follow-up. Unfortunately, the patient was lost to follow-up.

Discussion

Chiari malformation (CM) is a congenital disease, often identified in children and young adults, and first described by an Australian pathologist Hans Chiari [1]. He also classified CM into 4 subtypes CM 1-4 depending on the herniated structure and the degree of herniation. CM1 is a cerebellar tonsillar herniation below the foramen magnum greater than 5 mm. CM2 is a caudal descent of the vermis, medulla, and the fourth ventricle down the foramen magnum. CM3 is a herniation of the hindbrain structures into a posterior cranial encephalocele. CM4 is a hypoplastic or aplastic cerebellum in addition to tentorial hypoplasia. However, CM4 is no longer recognized [3]. The herniation of these parts through the foramen magnum is a source of different signs and symptoms such as headache [6], pain, weakness, sensory deficit, apnea, and others [1]. Later on, 2 more CM types were identified CM1.5 and CM0 [3].

CM0 subtype is used to describe a patient with CM symptoms with no cerebellar herniation or minimal, with syringomyelia. It is a rare subtype first described by Iskandar et al. [3,7]. CM0 and CM1 share the same presumed pathophysiology of abnormal cerebrospinal fluid circulation and a decrease in the size of posterior cranial fossa bones [7,8]. Syringomyelia, which is highly connected to CM1, was also linked to CM0 and played a major role in diagnosing it. However, it is not currently required to achieve the diagnosis as multiple cases of CM0 was reported without syringomyelia [4,5]. CM0, in rare cases, may be associated with syringobulbia, a cephalic extension of the syringomyelia [5].

MRI revolutionized CM diagnosis, where patients with or without symptoms were diagnosed and managed at earlier ages [6]. However, MRI findings do not necessarily correlate with symptoms [7]. Therefore, management of the patients should be based on the clinical presentation alongside the radiological findings [5,7]. Treatment of CM0 is the same as CM1, where it is targeted to decompress the posterior cranial fossa, which will alleviate the symptoms, supporting the previously mentioned theory on pathogenesis [5,7,8].

Conclusion

Chiari zero malformation is a rare subtype of Chiari malformation. It becomes even rarer with the association of syringobulbia. This case supports the literature with an extremely rare disease in an uncommon age group. More cases are being reported in the literature which may reveal new facts about the presentation and management of such cases.

Patient consent

An informed written consent was obtained from the patient for publishing this case report.

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