Case Report: Chiari Zero Malformation

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ABSTRACT

Introduction: Chiari zero, initially dubbed syringohydromyelia without hindbrain herniation, is a rare subset of Chiari malformations. The clinical presentation is usually due to syringohydromyelia. The mode of management is foramen magnum decompression. We report a case of Chiari 0 with multiseptated/multiloculated syringohydromyelia and review the literature. **Case report:** We presented a 41-year-old man with an 8-year history of progressive numbness. A neurological exam revealed left-sided upper-limb hypopselaphesia, diminished algesthesia, and temperature sensation in the left upper limb and trunk. MRI noted cervicothoracic syringohydromyelia with tonsillar herniation. We performed foramen magnum decompression with duraplasty. His syringomyelia reduced significantly, and his neurological status improved during the three-month follow-up. **Conclusion:** Chiari 0 is a rare entity thought to occur due to altered dynamics in CSF flow at the craniovertebral junction with resultant syringohydromyelia without tonsillar herniation. Foramen magnum decompression with duraplasty remains the most widely accepted surgical intervention.

Keywords: Chiari zero, Chiari-like malformation, syringomyelia, tight cisterna magna, atlantoaxial instability, foramen magnum decompression, atlantoaxial fusion

INTRODUCTION

Hindbrain malformations were initially described in 1829 [1] by Jean Cruveilhier. He reported a myelomeningocele patient in whom the dilated cervical region contained both the medulla oblongata and the cerebellum. [1]. Chiari described the four types of hindbrain malformations. [2-3] Since Chiari's original description, newer subtypes of Chiari malformation have been described, including Chiari zero malformation. Chiari zero malformation describes a subset of Chiari malformation characterized by syringomyelia with no evidence of hindbrain herniation that improves clinically or radiologically after posterior fossa decompression, usually indicating a functional disturbance of CSF flow across the craniovertebral junction. [4-5] We describe a patient with Chiari zero who underwent foramen magnum decompression with subsequent symptom resolution.

CASE REPORT

A 41-year-old man presented to the neurosurgical clinic with an eight-year history of progressive numbness on the left upper limb and trunk. The onset was acute, with progressive worsening beginning with the left hand and progressing to include the left upper limb and trunk.There was no history of trauma or any other events that could have explained the onset of symptoms.

A neurological examination revealed hypopselaphesia and complete loss of algesthesia in the left upper limb, as well as decreased algesthesia and temperature sensation in the left trunk. There was left-sided asymmetry associated with shoulder sagging. Deep sense, as well as proprioception and 2-point discrimination, were normal. All the limbs had normal natural myodynamia and muscle tension. An MRI of the whole spine showed multiseptated/multiloculated syringohydromyelia (C2-T6) and a shortened clivus. (Fig. 1A) There was no tonsillar ectopia or tethered spinal cord in the spine MR images. The posterior cranial fossa volume was 233 cc. The CT scan of the head was normal.

The patient underwent foramen magnum decompression with duraplasty and arachnoidal adhesiolysis. The left upper limb numbness and the hypopselaphesia improved, with the patient reporting significant changes two months post-decompression. We did not do atlantoaxial arthrodesis. The post-decompression MRI (Fig. 1B) showed thin residual syringohydromyelia compared to before surgery. We obtained written informed consent for the publication of this report and the accompanying images.

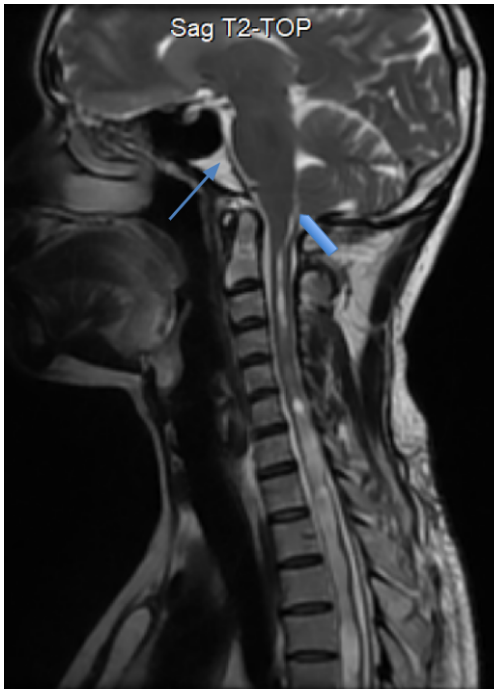

FIG 1. A Illustrates syringomyelia (C2-T6), shortened clivus (small arrow), with no tonsillar ectopia. Crowding at the FM (big arrow).

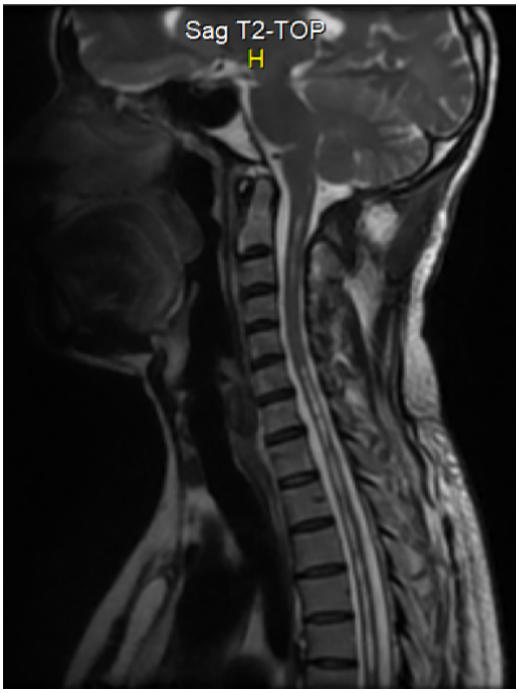

FIG 1B Post FMDD, showing thin residual syringohydromyelia, with suitable subarachnoid spaces at the level of the foramen magnum

CASE DISCUSSION

Hans Chiari described the four types of Chiari malformations. [2-3] New subsets have been developed due to clinical advancement. Eight classifications have been described: Chiari 0, Chiari I, Chiari 1.5, Chiari II, Chiari III, Chiari 3.5, Chiari IV, and Chiari V malformations. Chiari 0 and Chiari 1.5 have been used to describe the various subsets of Chiari 1. [6]

Chiari 0 is an infrequent finding and is thought to be a subset of Chiari 1 [7]. The most accepted theory of Chiari 0 malformations is that the alterations in CSF dynamics at the craniovertebral junction lead to the development of syringohydromyelia. Our patient had cervicothoracic syringomyelia. The presumptive mechanism for the genesis of these alterations is thought to be due to crowding at the foramen magnum, an arachnoid web, compression by a ligamentous band, or a "tight cisterna magna," where there is tonsillar compaction of CSF outflow above the foramen magnum, usually involving the foramen of Magendie. CSF flow could also be obstructed due to a distorted posterior fossa [4]. However, Goel [8-9] reported atlantoaxial instability as a significant driving force in Chiari malformation and syringomyelia.

The experts' inability to reach a consensus may lead to imprecise diagnostic criteria, causing incorrect CM0 diagnoses and unnecessary surgery. [17] Chiari 0 malformation has recently been described as: the obliteration of the cisterna magna and/or volumetrically small posterior cranial fossa, cerebellar tonsils positioned at the foramen magnum, and cervical syringomyelia. [17-18] The diagnosis requires careful patient selection after ruling out other causes of syringomyelia. [12] Our patient had cervicothoracic syringomyelia and obliteration of the cisterna magna by arachnoid adhesion, with the position of the tonsils noted to be at the level of the foramen magnum.

The prevalence of asymptomatic or symptomatic Chiari 0 malformations is unknown. As in our case, symptomatic patients with Chiari malformation have been reported to have a short clivus compared to asymptomatic patients. [19] Symptoms in Chiari 0 are usually because of syringohydromyelia or syringobulbia. Syringobulbia may occur in extreme cases. [7]. The most common symptoms in a patient with Chiari 0 have been reported to be headaches and neck pain. [19] This was not the case with our patient, since he presented with dissociated sensory loss. This is thought to be caused by disruptions in the decussating spinothalamic tracts, which result in loss of algesthesia and temperature. Light touch and proprioception were spared. [20] Scoliosis in patients with syringohydromyelia, as seen in this case, is thought to occur because of dysfunction in the anterior horn cell with resultant asymmetric weakness of the paraspinal muscles. [20] Syringomyelia's motor symptoms resemble those of central cord syndrome. [20] Bladder impairment often denotes a late finding. Other clinical findings include cranial nerve palsies, Brown-Sequard syndrome, atrophy of the musculature, and paraparesis.

MRI is the imaging modality of choice. [16] Standard MRI, both T1 and T2 weighted, with thin slices, usually not more than 3 mm, should be acquired. Imaging of the entire brain and spine should be undertaken to rule out significant cranial and spinal lesions. It is also essential to rule out evidence of high or

low intracranial pressure. CSF flow evaluation with cardiac-gated phase contrast (cine) CSF flow imaging helps to assess for disruptions of CSF flow and pulsation abnormalities at, above, or below the foramen magnum. [16] Flexion and extension x-rays or CT scans are vital in determining the presence or absence of atlantoaxial instability, which is critical in determining the choice of surgical procedure. [11] In our case, we did a whole spine MRI and head CT scan.

Foramen magnum decompression remains the gold standard surgical management procedure. [4] This may or may not include duraplasty, partial dural opening, tonsillar coagulation, and removal of arachnoidal bands. Most literature [10] advocates choosing procedures that effectively expand the volume of the foramen magnum beyond bony decompression alone, either selecting simple craniocervical decompression and duraplasty without opening the arachnoid membrane or decompression of the bone of the posterior lip of the foramen magnum and removal of the posterior arch of C1, and occasionally part of C2, with a partial thickness dural incision [10]. Goel [9,11], however, proposed atlantoaxial stabilization without foramen magnum decompression as a blanket solution to Chiari 1 malformations and its subsets, including Chiari zero. We opted to do decompression, duraplasty, and arachnoid adhesiolysis. The failure rate of foramen magnum decompression surgery in major surgical series ranges from 15% to 50%. [13-15]

CONCLUSION

Chiari 0 is a rare entity thought to occur due to altered dynamics in CSF flow at the craniovertebral junction with resultant syringohydromyelia without tonsillar herniation. Foramen magnum decompression with duraplasty remains the most widely accepted surgical intervention.

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