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Syringobulbia and syringomyelia in a case with Chiari 0 malformation successfully treated by posterior fossa reconstruction. Case presentation and literature review

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Chiari zero malformation, syryngomyelia, syringobulbia, cerebellar tonsils

ABSTRACT

"Chiari zero malformation" is a rare and a relatively new described condition which associates syringohydromyelia without caudal displacement of the cerebellar tonsils through the foramen magnum. We present a case of a 40 years old woman with Chiari zero malformation with both syringomyelia and syringobulbia and a good clinical and radiological outcome after posterior fossa decompression. The presence of associated syringomyelia and syringobulbia in this condition is less frequent and it usually occurs in younger patients. In our case we considered syringobulbia as being an extension of syringomyelia.

INTRODUCTION

The Chiari malformations were first described by Hans Chiari in terms of the cerebellar tonsils herniating out of the skull and he identified four different types, of which the type I and II are more common (4). Nowadays, most people define Chiari I as tonsillar herniation of greater than 3-5 mm, measured as the distance below the foramen magnum and the condition is often associated with syryngomyelia (1). But patients with syringohydromyelia without hindbrain herniation that respond to posterior fossa decompression have been described and the condition was named "Chiari zero malformation" (7). Conversely, many patients can have herniations greater than 3-5 mm, but with no symptoms, so tonsillar herniation identified radiographically is of limited prognostic value and requires clinical correlation (5). The association with syringobulbia is even a rarer situation, with only a few cases in the literature and especially in children (19,20). Hereby we present a case of a women with Chiari zero malformation and the presence of both syringomyelia and syringobulbia who was treated surgically in our department.

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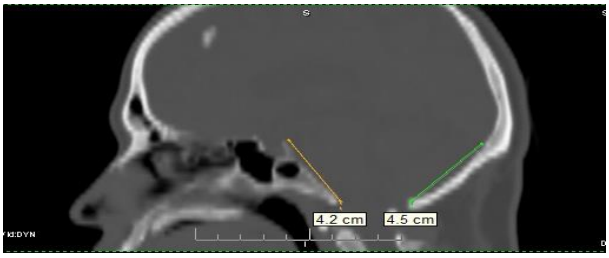
Case report

A 40-year-old woman was admitted to our hospital with a 2-years history of progressive proximal left-sided upper limb weakness. At admission she had a decreased sensitivity to painful stimuli associated with decreased temperature sensation as well in the upper limbs and swallowing difficulties. She also presented kypho-scoliosis developed during the childhood.

The patient was investigated with a head computed tomography (CT) and a 1.5T IRM of the cranio-cervical junction, cervical and thoracic spine.
On the preoperative images, we made some linear measurements for posterior fossa dimensions: the length of the clivus defined as the distance from the top of the dorsum sellae to the basion, and the length of the supraocciput measured between the internal occipital protuberance and the opisthion. We also calculated the spheroidal posterior fossa volume (PFV) based on the spheroidal formula (6):
PFV=4/3 x π x (X/2 x Y/2 x Z/2), where: X is the anteroposterior measurement from the posterior clinoid process to the torcula; Y is the height of the posterior fossa measured from the basion to the peak of the tentorium cerebelli; and Z is the maximum width of the posterior fossa. (6)

To assess the syrinx characteristics, we noted the cranial and caudal extent and we measured on the pre and postoperative images the ratio between the syrinx and the spinal cord diameter at the level of the maximum expansion. In addition, we calculated the ratio of the surface area of syrinx to spinal cord using the formula πr^2 for surface area, with r being the radius (7).

The clival length of our patient was 4,2 cm and the length of the supraocciput was 4,5 cm. The width of the posterior cranial fossa was 9,9 cm (Z=9,9 cm), and the anterior-posterior dimension was 8,3 cm (X=8,3 cm). The height from the basion to tentorium cerebelli was 5,3 cm (Y=5,3 cm). The calculated spheroidal posterior fossa volume was 227,9 cm^3. These measurements are consistent with a small posterior fosa.



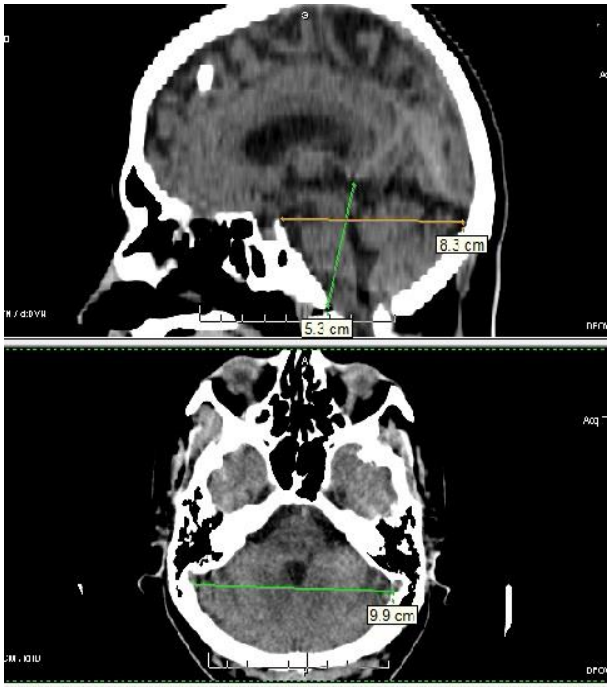
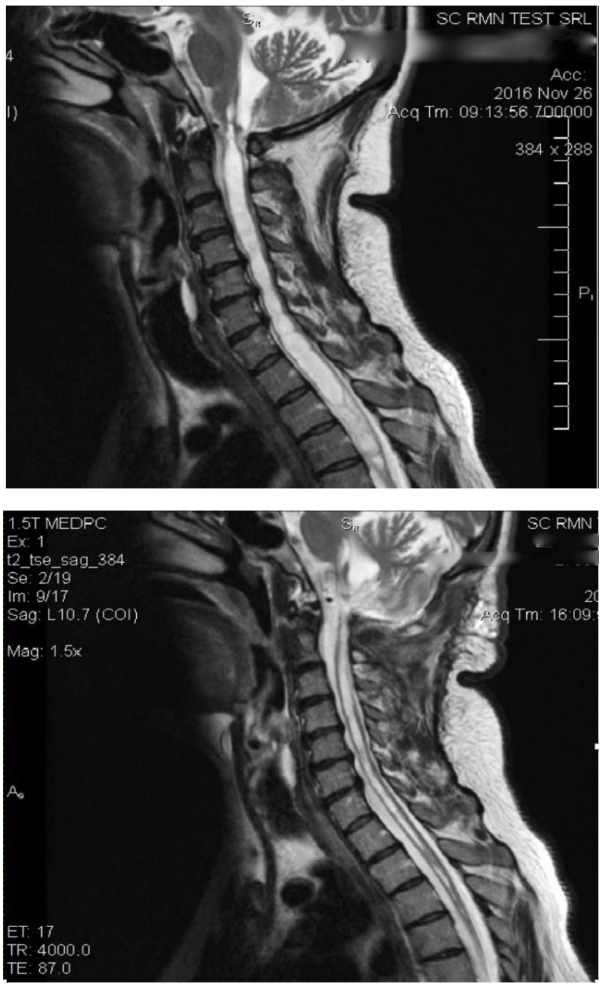


Figure 1. Posterior fossa measurements

Figure 2. Pre and post-operatively MRI images



The cranial extent of the syrinx was at the level of the medulla oblongata and the caudal extent was at the level of T4 vertebral body. We measured the maximum expansion of the syrinx cavity at the level of the T1 vertebral body and compared the pre and 3 months postop values. The ratio of the preoperative syrinx/spinal cord diameter at the level of greatest expansion (T1 vertebral body) was 0.81 (9mm/11mm). The ratio of the postoperative syrinx/spinal cord diameter at the same vertebral level was 0.37 (3mm/8mm). The ratio of the preoperative syrinx/spinal cord surface at the level of greatest expansion (T1 vertebral body) was 0.67, and 3 months postoperatively was 0.14

The patient was surgically treated by performing a craniocervical decompression which consisted of a suboccipital craniectomy, C1 laminectomy, intradural lysis of the arachnoidal adhesions that occluded the foramen of Magendie and duraplasty. She had a good post-operative evolution with the progressive remission of the symptoms.

Discussion

Chiari 0 malformation is a rare and a relatively new described condition which associates syringohydromyelia without caudal displacement of the cerebellar tonsils through the foramen magnum, of which clinical symptoms and radiological aspect can be ameliorated after posterior fossa decompression. (7,18). While Chiari II and III malformations can be explained by a maldevelopment of the posterior fossa neural structures, more and more evidence point to an underdevelopment of the bony structures of posterior fossa, leading to a smaller p-fossa that contain a normal developed neural tissue as an explanation for Chiari I and Chiari 0 malformations (2,8,10,11,12,13,14,15,16).

Syringomyelia may be associated with many conditions such as Chiari malformations, spinal trauma, spinal tumours, tethered cord or spinal dysraphism. In a study of Strahle et al, the authors identified a number of 271 patients with a syrinx out of 14118 patients who undergone brain or spine imaging during a period of 11 years. The Chiari malformation type I was the most common associated condition with syringomyelia and the syrinx was more likely to have the cranial extent in the cervical spine and to have a width greater than 5 mm compared with other conditions. They found only 4 patients with Chiari 0 malformation but the syrinx in these cases had the big-gest width and length, bigger than syrinx associated with Chiari type I and II malformations. Al-so the Chiari type 0 patients had a more superior cranial extent of their syrinx. (17).

We considered that the characteristics of our patient are consistent with the Chiari 0 malformation. The measurements corresponded with a small posterior fossa and the MRI revealed syringomyelobulbia without tonsillar herniation. Three months postoperatively, the symptomatology improved, the algesthesia and temperature sensation in the upper limbs were better than preoperatively, the swallowing difficulties disappeared and the abduction of both upper limbs obviously improved. The MRI performed 3 months postop showed that syringomyelia diminished semnificatively compared with that before surgery.

Conclusion

The presence of associated syringobulbia with syringomyelia in the Chiari zero malformation is a condition rarely described in the literature and in the most of the cases the patients are younger. (19,20). In our case we considered syringobulbia as being an extension of syringomyelia.

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