Case Report

Resolution of Syringomyelia Following VP-shunt in Chiari-I Malformation: A Case Report

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Abstract:
Indications for surgery and the surgical technique of foramen magnum decompression for patients with Chiari I malformation and syringomyelia are controversial issues. This case report supports the view that patient with syringomyelia who underwent VP-shunt and subsequent observation may be adequate for patients without progressive symptoms or with mild clinical symptoms.

Key words: Chiari-I malformation, Syringomyelia, V-P shunt, Resolution.


Introduction:
Obstruction of cerebrospinal fluid (CSF) flow due to herniation of cerebellar tonsils into or even below the level of the foramen magnum (i.e., Chiari I malformation) is one of the most common causes of syringomyelia. A herniation of more than 5 mm is considered pathological for adults.¹ Studies have shown that about 75 to 85% of patients with Chiari I malformation develop syringomyelia.²-⁴ Chiari I malformation may develop as a congenital disorder of the mesoderm because of a small posterior fossa.⁴-⁶ A second group of patients may acquire Chiari I malformation because of lumboperitoneal shunts,⁷ tumors of the posterior fossa,⁸ birth trauma,⁹ or meningeal reactions at the foramen magnum.¹⁰ The natural history of clinical symptoms in this anomaly is not well known. No general agreement exists with regard to which patients should have surgery. Should every patient with Chiari I malformation undergo surgery? Should surgery be postponed until symptoms start to develop? In this article, we report on an adult patient who experienced spontaneous resolution of Chiari I malformation and syringomyelia during a period of 3 months following VP-shunt.

Case report:
A 22 years old male presented with a 3 months history of numbness and tingling sensation of left hand and left foot. Neurological examination revealed reduced muscle power in left arm Medical Research Council (MRC) grade 4(-). Other remaining examinations were normal, with no long-tract signs. A magnetic resonance imaging (MRI) scan showed a classic Chiari I malformation with syringomyelia between C2 to D4 (Fig. 1). The syrinx affected the left hemicord corresponding to the clinical findings. Obstructive hydrocephalus was present (Fig. 3). The advantages and risks of surgery were discussed with the patient. Ventriculo-peritoneal shunt was placed. Within the next couple of months, symptoms started to regress...
spontaneously, and surgery was therefore postponed. Two months later, the patient was asymptomatic and underwent a second MRI examination, which showed resolution of the Chiari I malformation and syringomyelia (Fig. 2). The patient did not recall any specific incidents associated with sudden changes of symptoms in these 2 months but mentioned a gradual improvement in muscle power.

Discussion:
Moriwaka et al.\textsuperscript{11} published a nationwide epidemiological survey on syringomyelia in Japan. Among 1243 patients with syringomyelia, 684 (51.2\%) cases had associated Chiari I malformation. The clinical course was slowly progressive in the great majority of patients; in 202 patients (16.2\%), however, a rather stable course was observed, and

\textbf{Fig.-1:} Sagittal -T2-weighted MRI scan showing Chiari I malformation with the descend of tonsils into the foramen magnum to the upper limit of C1, occluding the cisterna magna and a large syrinx starting at the level of C2 to D4.

\textbf{Fig.-2:} T2-weighted MRI scan of the same patient, obtained 2 months after presentation which shows the regression of the tonsils into the posterior fossa, a normal-appearing cisterna magna, and partial resolution of the syrinx.

\textbf{Fig.-3:} Axial -T1-weighted MRI scan showing features of obstructive hydrocephalus.
spontaneous resolution of symptoms was observed in only 29 patients (2.3%). Similar figures were reported for German patients by Schliep and Ritter in 1971 (60 patients) and by Hertel et al. in 1973 (323 patients). The number of 664 patients with spontaneous clinical improvement who also demonstrated radiological resolution of syringomyelia and/or tonsillar herniation was not examined in any of these reports.

To date, very few cases of spontaneous resolution of syringomyelia in patients with Chiari I malformation have been documented accurately with MRI. To our knowledge, only 10 patients similar to our patient have been reported. Another two patients with syringomyelia that was unrelated to Chiari I malformation also showed spontaneous resolution of the syrinx. Among the 10 reported patients with Chiari I malformation, 5 were adults and 5 were children.

Only five presented with neurological symptoms; the other five were asymptomatic. In all but one patient, symptoms regressed together with the syrinx. In three of the pediatric patients, a remarkable improvement of the Chiari I malformations was observed; in the other two the Chiari I malformations disappeared completely. In one patient, however, the Chiari I malformation and syringomyelia reappeared without accompanying neurological symptoms. Evidence is mounting that syringomyelia develops in association with obstruction of CSF flow at the foramen magnum in Chiari I malformation. Therefore, spontaneous resolution of a syrinx requires spontaneous improvement in CSF flow. In children, the growth of the cranium continues after the cerebellum has reached more than 90% of its final size by age 2 years. Therefore, the tonsils may regress intracranially in relation to the growing cranium to the extent that free CSF passage is established, thus allowing the syrinx to decrease in size or even disappear completely.29

A second possible mechanism involves changes in venous outflow in the posterior fossa. Girard et al. demonstrated that Chiari I malformation is a regular feature in children with vein of Galen aneurysms due to veno-occlusive disease. After successful embolization of the venous malformation, the Chiari I malformation and syringomyelia—if present—disappeared. One may speculate that venous thrombosis involving the posterior fossa may cause descent of the cerebellar tonsils into the foramen magnum and that, after recanalization or establishment of sufficient collateral flow, the tonsils may regress intracranially. The MRI scans of the patient described here, however, did not show evidence of cerebellar swelling, which one would expect if venous thrombosis were present. Furthermore, venous thrombosis capable of producing tonsillar herniation generally is associated with more pronounced neurological symptoms. Aside from children with vein of Galen aneurysms, no patients with documented venous thrombosis or partial venous obstruction causing Chiari I malformation have been described. Another possible mechanism of spontaneous resolution involves supratentorial lesions such as hydrocephalus or other space-occupying processes. This possibility was excluded in this patient by an examination of the patient’s cranial MRI scans. Among six adult patients, including the one reported in this article, significant improvement of Chiari I malformation accompanied the resolution of syringomyelia in three patients, and Chiari I malformation remained unchanged in the other three patients.

A changed ratio between cerebellar and intracranial volumes cannot explain the disappearance of CSF flow obstruction and syringomyelia in adults; something else must have happened. Jack et al. and Santoro et al. proposed an interesting mechanism by which these symptoms spontaneously resolve. They suggested that with vigorous Valsalva maneuvers, the pressure inside the syrinx may increase to an extent that CSF tears the spinal cord, thus creating a communication between the cavity and the spinal subarachnoid space, which subsequently drains the cavity. Such communications between syrinx and subarachnoid space have been demonstrated by MRI studies and neuropathological examinations. Even though this phenomenon may explain the decrease of a syrinx cavity in some instances, communications between syrinx and subarachnoid space are so common that spontaneous resolution of syrinx cavities would be observed much more often if this circumstance were the decisive factor. Furthermore, this mechanism would not explain the disappearance of the Chiari I malformation. The pathogenesis of syringomyelia associated with Chiari I malformation still is not fully understood.
Gardner and Angel’s hydrodynamic theory [32] and Williams’s hypothesis of cranio cervical pressure dissociation[27] postulate communication between the fourth ventricle and the syrinx via the obex and the central canal. Neuropathological studies [33-35] have shown that such communication does not exist in the overwhelming majority of patients. Cardiac-gated cine-MRI studies have shown that obstruction of CSF flow is always present in patients with Chiari I malformation and abnormalities associated with syringomyelia.[2,24]

Surgical procedures that establish a significant improvement in or even normalization of CSF flow lead to a decrease in or even disappearance of syringomyelia. In patients with Chiari I malformation, CSF flow obstruction at the foramen magnum has two possible causes: 1) the subarachnoid space may be obstructed by tonsillar tissue, and/or 2) arachnoid scarring may occlude subarachnoid pathways. Arachnoid changes at the foramen magnum and foramen of Magendie are much more common in patients with Chiari I malformation than is generally acknowledged and may contribute to CSF flow obstruction and neurological symptoms.[2] If the spontaneous resolution of a syrinx requires the spontaneous resolution of CSF flow obstruction, the most likely explanation for the resolution of symptoms in the patient presented here is a spontaneous rupture of an arachnoid membrane that improved the passage of CSF at the foramen magnum to such an extent that the syrinx decreased in size. Might Chiari I malformations improve with normalization of CSF flow at the foramen magnum? Several neuroradiological studies of patients who had surgery for Chiari I malformations demonstrated that the cerebellar tonsils retract intracranially, provided that a decompression of the foramen magnum is performed with formation of a cisterna magna and opening of obstructed CSF pathways.[25,26]

Conclusion:
Spontaneous resolution of syringomyelia and Chiari I malformation is exceptionally rare and may be observed only if CSF flow obstruction resolves spontaneously. Therefore, we recommend surgery for Chiari I malformation in adult patients with neurological symptoms. In young children, a period of observation is warranted. If the tonsils regress together with the child’s continuing cranial growth, surgery may not be required. If the child’s symptoms persist or even progress, however, surgery should be recommended. Surgery in these patients should serve two purposes: the decompression of central nervous structures at the foramen magnum and the opening of CSF pathways to ensure normal CSF flow.

References:
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Mohammad Hossain et al


