

Syringomyelia regression after shunting of a trapped fourth ventricle

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Abstract

We describe a case of progressive syringomyelia following post-infectious trapped fourth ventricle (TFV), which resolved after shunting of the fourth ventricle. A 28-year-old female who had previously undergone treatment of intracerebral hemorrhage and meningitis developed a hydrocephalus with TFV. After 3 years she developed disturbance of walking and coordination. Cranial-CT revealed an enlargement of the shunted fourth ventricle as a result of shunt dysfunction. Furthermore a cervical syringomyelia developed. The patient underwent a revision of a failed fourth ventriculo-peritoneal shunt. Postoperatively, syringomyelia resolved within 6 months and the associated neurological deficits improved significantly. An insufficiency of cerebrospinal fluid draining among patients with TFV can be associated with communicating syringomyelia. An early detection and treatment seems important on resolving syringomyelia and avoiding permanent neurological deficits. Ventriculo-peritoneal shunt in trapped fourth ventricles can resolve a secondary syringomyelia.

Introduction

Trapped fourth ventricle is a rare and uncommon entity which has been observed as a complication after intraventricular hemorrhage, infection/meningitis or as a result of chronic over drainage after hydrocephalic shunting.^{1,4} A post-infectious occlusion of fourth ventricle outflow (foramina of Luschka and Magendie) and sylvian aqueduct is the second most common cause for the development of trapped fourth ventricle.⁵ An increased cerebrospinal fluid (CSF) pressure within the fourth ventricle can lead secondary to the enlargement of the central canal in terms of communicating secondary syringomyelia. The exact pathophysiological mechanism of developing syringomyelia generally is not well established and remains yet controversial although several theories have been postulated.

We report a case where a post-infectiously trapped fourth ventricle leads to syringomyelia.

Case Report

A 28-year-old female was admitted in our department because of a large right-sided intracerebral hemorrhage as a result of a ruptured arteriovenous malformation. She underwent neurosurgical and endovascular treatment. Due to a post hemorrhagic hydrocephalus external ventricular drain was performed. During the intensive care treatment she unfortunately developed meningitis. After antibiotic treatment, the meningitis was cured and a ventriculo peritoneal shunt was performed. However, the follow up imaging, revealed an isolated enlarged fourth ventricle despite shunting of the lateral ventricles. Therefore a shunting of the fourth ventricle has been done as well. Nearly 22 months later the patient presented again with secondary walking and coordination impairment.

Neurological examination showed increasing ataxia and eye movement disorder beside the known left-sided hemiparesis. Cranial computed tomography (CT) demonstrated remarkable dilation of fourth ventricle indicating a shunt dysfunction. Furthermore syringomyelia without tonsil herniation has occurred, which was identified through cervical magnetic resonance imaging (MRI) extending from C2 to C7. The complete replacement of the dysfunctional shunt system was immediately performed. Postoperatively, the syringomyelia resolved within 6 months and the associated neurological deficits improved as well. An entire resolution of the trapped fourth ventricle was confirmed on CT scan (Figure 1).

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Discussion

The fact that acquired lesions in region of the foramen magnum like Chiari malformation, hydrocephalus, tumors of the posterior fossa posterior or CSF-over drainage are associated with tonsil herniation leading to a secondary syringomyelia is well established.⁶

The pathophysiology of syrinx formation still remains unclear. However, pressure difference between the cranial cavity and the spinal central canal causes CSF flow disturbance at

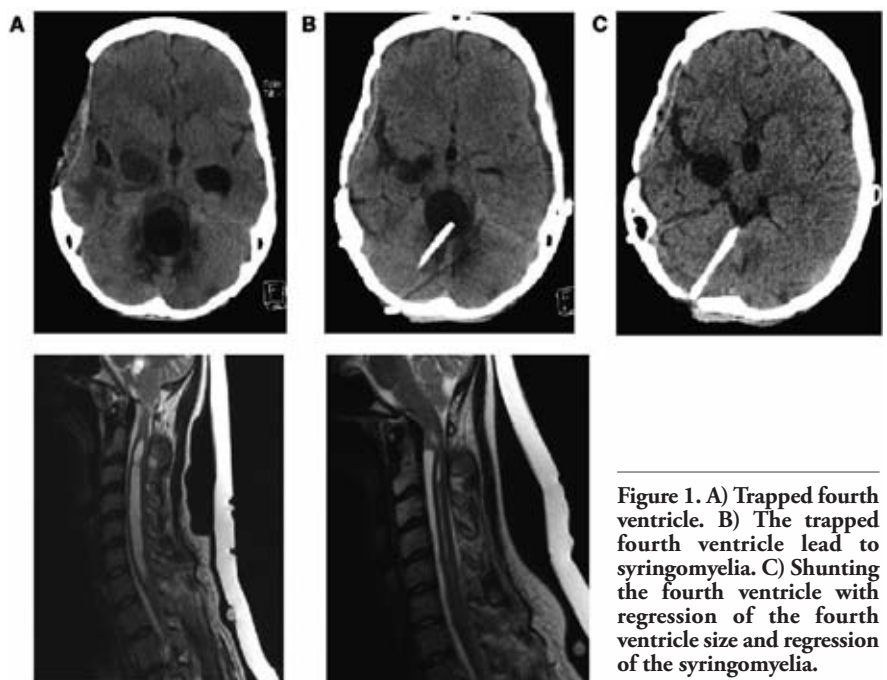


Figure 1. A) Trapped fourth ventricle. B) The trapped fourth ventricle lead to syringomyelia. C) Shunting the fourth ventricle with regression of the fourth ventricle size and regression of the syringomyelia.

the cranio-cervical junction seems to play a crucial role. Initially, it was hypothesized that cardiac systolic pulsations are directly transmitted from the intracranial cavity to the central canal inducing syrinx formation in terms of a hydrodynamic *water-hammer*.⁷ The use of diagnostic MRI disproved this theory since it has been shown that communication between the fourth ventricle and a patent central canal is not required for the formation of a syrinx.

Another explanation is that acquired lesions at the cranio-cervical junction can occlude the pulsatile flow of CSF along the subarachnoid space the cranio-cervical region causing an increased CSF-pressure gradient from the perimedullary subarachnoid space to the spinal cord. This causes an increase of fluid within the central canal creating a syrinx.⁸

In our case meningitis with basilar arachnoiditis led to an occlusion of the fourth ventricular outflow. A Secondary progressive syrinx developed.

Treatment extends from placement of fourth ventriculo-peritoneal shunt, endoscopic aqueductoplasty and interventriculostomy to open fenestration via suboccipital craniotomy.^{9,10} We consider that any of these options has their assets and drawbacks so the choice of treatment should be individually taken depending on patient history and pathophysiology. The purpose of this report is to illustrate the possibility of syrinx resolution/treatment (caused by a trapped fourth ventricle) with the drainage of the fourth ventricle via a ventriculo-peritoneal shunt. Implantation of a fourth ventriculo-peritoneal shunt

proved as a simple, feasible and effective treatment modality.

In cases of syringomyelia caused by Chiari malformation the therapeutic strategy is different than that chosen in the present paper. In Chiari I malformations and syringomyelia in general, Isu *et al.* recommended ventriculo-peritoneal shunting, whereas Klekamp *et al.* suggest small craniectomy with dural opening and arachnoids dissection.^{11,12}

In our case there was no compression of the fourth ventricle caused by stenosis in the foramen magnum, therefore we decided to treat only with shunting.

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