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Case Report

Treatment of delayed Chiari malformation and syringomyelia after lumboperitoneal shunt placement: Case report and treatment recommendations

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Abstract

Background: Delayed Chiari malformation with syringomyelia is an uncommon and sparsely reported complication after lumbar cisternal shunting. A number of treatments have been implemented with varying degrees of success. After review of these modalities, a treatment plan was devised and implemented.

Case Description: A 15-year-old girl was diagnosed with idiopathic intracranial hypertension (pseudotumor cerebri) and had placement of a Medtronic Spetzler lumboperitoneal shunt. She did well for 10 years and then developed a 3-week history of headache, balance problems, and sensory loss. Workup demonstrated a new Chiari malformation and cervical syrinx. She underwent initial placement of a ventriculoperitoneal shunt, followed by ligation of the lumboperitoneal shunt. She then had a suboccipital decompression and duraplasty. Follow-up at 6 months showed relief of her symptoms and MRI demonstrated resolution of the Chiari malformation and syrinx.

Conclusion: Delayed Chiari malformation and/or syringomyelia after lumbar shunting is best treated with placement of a ventricular device and ligation of the lumbar shunt. The Chiari malformation and/or syrinx should be surgically addressed for persistent symptoms or progression of the structural abnormality.

Key Words: Acquired chiari malformation, idiopathic intracranial hypertension, lumboperitoneal shunt, pseudotumor cerebri, syringomyelia

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INTRODUCTION

First recognized in the late 1970s and 1980s, a sparsely reported delayed complication of lumbar cisternal shunting is the formation of an "acquired" Chiari I malformation with or without the formation of a syrinx.[4,6,12,13] Since then, a number of cases have been reported with a variety of treatment techniques. These techniques include cranial decompression,[2,6] placement of a lumbar shunt valve,[11] placement of a ventricular peritoneal shunt, and ligation of the lumboperitoneal (LP) shunt with or without suboccipital decompression;[4,7,12,13] and in cases of a syrinx only, drainage of the syrinx.[7] This report details a case of delayed acquired Chiari malformation and syringomyelia after LP shunting for pseudotumor cerebri, and our treatment rationale with recommendations.

CASE REPORT

An overweight 15-year-old girl developed severe headache and visual loss with papilledema. A cranial magnetic resonance imaging (MRI) scan, MR angiogram, and MR venogram were negative. Serial lumbar punctures revealed elevated opening pressures consistent with idiopathic intracranial hypertension, i.e. pseudotumor cerebri [Figure 1]. Refractory to medical treatment, she underwent placement of a Medtronic Spetzler LP shunt. The Spetzler shunt has slit valves at its distal end and a catheter of small diameter, which provide resistance to CSF flow in order to regulate pressure. The patient did well with resolution of symptoms for 10 years.

At age 25, she developed a 3-week history of severe headache and intractable nausea and vomiting. Vision remained intact as did her motor exam. However, decreased touch and pinprick sensation were noted in the left upper extremity. Repeat MRI of the head and cervical spine demonstrated an acquired Chiari I malformation with cerebellar tonsillar ectopia extending 16 mm below the foramen magnum, and a cervical syrinx from C2 extending to the T8 level [Figure 2]. Neither the Chiari I malformation nor the cervical syrinx were present on previous scans.

We assumed that the new findings were the result of cranial spinal CSF pressure differentials caused by the lumbar drain. Weight reduction can result in resolution of pseudotumor cerebri. Because the patient had not undergone substantial weight loss and was not experiencing papilledema, it was assumed that the pseudotumor cerebri continued to be managed by her lumbar drain and that the drain was still working. The patient underwent placement of a right frontal programmable ventricular peritoneal shunt utilizing stereotactic navigation and ligation of the LP shunt. Postoperatively, she continued to have severe headaches, nausea, and vomiting. Follow-up computed tomography (CT) showed decreased ventricular size and persistent Chiari malformation. Secondary to persistent symptoms, a suboccipital craniectomy, C1 laminectomy, and duraplasty were performed 5 days after shunting. Bipolar cautery was used to shrink the tonsils and clear egress of CSF from the fourth ventricle was noted prior to closure. With shrinkage of the ventricles, there appeared to be excessive ventricular tubing, so the proximal catheter was repositioned. The patient's symptoms gradually resolved over time, including the sensory symptoms in her left arm. A follow-up MRI done 6 months postoperatively revealed a complete resolution of both the Chiari malformation and the syrinx [Figure 3].

DISCUSSION

First recognized in the late 1970s and 1980s, acquired Chiari malformation and/or syringomyelia as a delayed symptomatic complication of lumbar cisternal shunting has been reported only sporadically.[4,6,12,13] Johnston *et al.*[7] reported an incidence of a new Chiari malformation in 11 of 70 patients treated with LP shunting for pseudotumor cerebri. Of these, eight were asymptomatic and three required treatment. Acquired Chiari malformation after lumbar cisternal shunting may be more prevalent in the pediatric population. In a retrospective study, Chumas *et al.*[1] noted tonsillar ectopia in 70% of patients, but less than 5% required surgical intervention. In a smaller series of 10 patients, 7 were found to have delayed tonsillar migration with 4 of the 7 undergoing surgery for symptoms.[10]

Delayed Chiari malformation and/or syringomyelia have been noted after lumbar cisternal shunting regardless of the underlying etiology. It has been described in cases of pseudotumor cerebri, shunting of cysts, and communicating hydrocephalus. It has been described

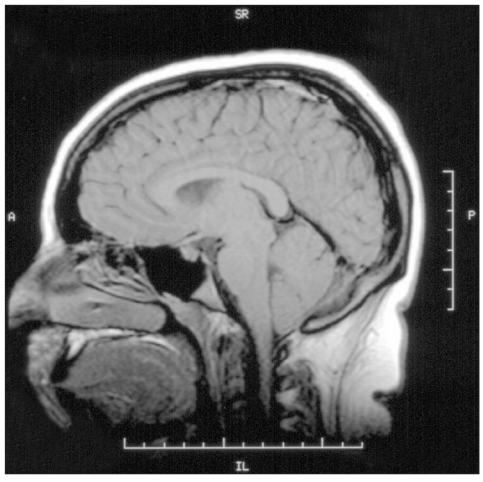

Figure 1: Normal sagittal view of the patient's cranial MRI at initial diagnosis of pseudotumor cerebri

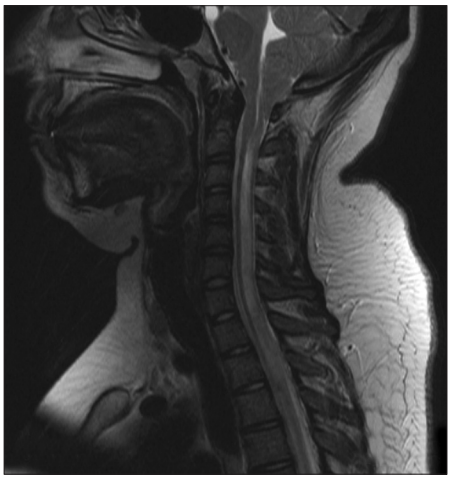

Figure 2: Sagittal cervical MRI of the same patient 10 years post lumboperitoneal shunting demonstrating a new "acquired" Chiari malformation and cervical syrinx

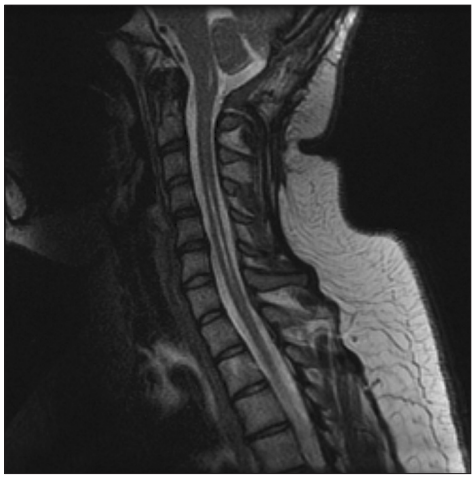

Figure 3: Sagittal cervical MRI 6 months after ligation of the lumbar shunt, placement of a ventricular shunt, and suboccipital decompression with resolution of the syrinx and Chiari malformation

in lumbar shunts to the peritoneum or ureter.[4] While the cause of the acquired Chiari malformation or syrinx is debatable, most surgeons feel it is a result of CSF pressure differentials across the cranial cervical junction, created by the drainage.[8,9,14]

A number of treatment modalities have been employed to resolve these complications, with varying degrees of success. For some patients, a cranial enlargement was successful without manipulation of the lumbar shunt.[2,6] In another case, a valveless system was replaced with a valve on the lumbar shunt with symptomatic improvement.[11] For cases of isolated Chiari malformation, a suboccipital decompression was performed with placement of a valve in the LP shunt in three patients with one failing.[7] When suboccipital decompression alone was attempted without adjusting the LP shunt in one case, the patient went on to develop a syrinx requiring ligation of the lumbar drainage and placement of a ventricular device with good results.[3] Four patients with isolated syringomyelia and without tonsillar ectopia were treated with syrinx shunting or syringotomy. Two of these four patients developed subsequent Chiari malformations and required surgical decompression.[7]

In a number of reports, the lumbar shunt was ligated and replaced with a ventricular atrial or peritoneal shunt without suboccipital decompression.[4,5,7,12,13] Symptoms resolved for many of these patients, but some continued to have issues requiring further surgery.[5,7] It should also be noted that while symptoms may improve with ventricular peritoneal shunting only, the structural abnormalities such as a syrinx do not always resolve.[10]

Placing ventriculoperitoneal shunts in pseudotumor cerebri patients is technically challenging and shunts may have a higher failure rate as the ventricles are usually small.

A review of the various treatments for acquired Chiari malformations and/or syringomyelia after lumbar shunting in these reports suggests that the greatest treatment success occurs with ligation of the lumbar drainage and placement of a ventricular device. This is a logical result if it is assumed that alterations in the cranial spinal fluid pressures across the foramen magnum are responsible.[14] Replacing the lumbar drainage with a ventricular shunt should concurrently eliminate this issue and continue to treat the underlying elevated CSF pressures. Shunting alone, however, may not reverse the structural abnormalities of a Chiari malformation or syrinx. Such abnormalities can still cause symptoms or even progress resulting in the need for surgical intervention. In our case, we chose to first ligate the lumbar shunt and place a programmable ventricular peritoneal shunt. Because the patient continued to have severe headaches, nausea, and vomiting, we proceeded to a suboccipital decompression. This ultimately resulted in resolution of both symptoms and the structural abnormalities.

In conclusion, acquired Chiari malformation and/or syringomyelia as a delayed complication after lumbar cisternal shunting occurs with some frequency, particularly in the pediatric population, but is not always symptomatic. Our review of these reported cases suggest the most effective treatment option is to remove the responsible factor of cranial cervical pressure differentials by replacing the lumbar shunt with a ventricular device. This procedure is made more feasible even with smaller ventricles using a stereotactic navigation device. Even if ventricular shunting is successful in alleviating symptoms, the patient should be monitored. Shunts alone may fail to resolve the Chiari malformation or syrinx, which may still progress to cause symptoms requiring treatment. Therefore, we suggest a low threshold for suboccipital decompression if symptoms persist or the structural abnormalities worsen.

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