



Case Report

Usefulness of cyst-subarachnoid shunt using syringo-subarachnoid shunt tube for symptomatic enlarging ventriculus terminalis: A case report and review of the literature

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ABSTRACT

Background: The ventriculus terminalis (VT) is a cystic embryological remnant of the conus medullaris that usually regresses after birth. This structure rarely persists into adulthood and may produce neurological symptoms. We recently encountered three cases of symptomatic enlarging VT.

Case Description: The three female patients were 78, 64, and 67 years old. Symptoms included pain, numbness, motor weakness, and frequent urination that gradually worsened. Magnetic resonance imaging revealed cystic dilations of slow growing VT. These patients showed marked improvement after cyst-subarachnoid shunt using a syringo-subarachnoid shunt tube.

Conclusion: Symptomatic enlarging VT is an extremely rare cause of conus medullaris syndrome and the optimal treatment strategy remains unclear. Surgical management may thus be appropriate for patients with symptomatic enlarging VT.

Keywords: Cyst-subarachnoid shunt, Enlarging ventriculus terminalis, Slow growing, Symptomatic case, Syringo-subarachnoid shunt tube

INTRODUCTION

The ventriculus terminalis (VT), also known as the fifth ventricle, is a small ependymal-lined fusiform dilatation of the terminal central canal of the spinal cord, located at the transition from the tip of the conus medullaris to the origin of the filum terminale.^[2-4,10] The VT is present in humans during fetal development and usually regresses completely after birth. However, persistence into adulthood represents a rare pathology with an unclear pathogenesis. Even then, VT is generally asymptomatic and rarely requires treatment because the appearance of symptoms as the cyst enlarges is extremely rare.^[1,3,10] As a result, treatment strategies for this condition have not been established. Here, we describe three cases of symptomatic, enlarging VT in adulthood and propose the utility of cyst-subarachnoid shunt using a syringo-subarachnoid shunt tube for symptomatic enlarging VT.

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CASE DESCRIPTION

The clinical study of the following cases was approved by the Ethics Committee for Clinical Research at Ehime University Hospital, and informed consent was obtained from the patients before initiating the study.

Case 1

A 78-year-old woman with no contributory history of illness or medical treatment was admitted to our hospital with low back pain radiating to both limbs, numbness in both soles, intermittent claudication, and frequent urination. No paralysis was apparent in either lower limb. Evaluation of the superficial and deep tendon reflexes did not demonstrate any abnormality. She had been asymptomatic until 12 years earlier. Magnetic resonance imaging (MRI) revealed the presence of a cystic lesion located in the conus medullaris, with a well-defined outline and no enhancement on gadolinium-enhanced T1-weighted imaging, confirming cystic dilation of the VT as so-called enlarging VT. The size of this cyst appeared to have increased over the 12 years [Figures 1a and b]. As a result, we determined that this condition represented symptomatic enlarging VT and planned a surgical intervention. The patient underwent a T11-T12 laminectomy. On opening the dura, the expanded conus began herniating outward under pressure, which was relieved by fenestration. Cerebrospinal fluid-colored liquid was drained to the subarachnoid space and a cyst-subarachnoid shunt was created using a syringo-subarachnoid shunt tube (diameter, 1.2 mm; length, 40 mm; Createmedic Co). Although neurological symptoms improved markedly

after the surgical intervention, the patient reported residual numbness slightly in the left hallux. Follow-up MRI after 3 years revealed complete regression of the dilation with no evidence of relapse [Figures 1c and 2]. No recurrence of neurological symptoms has been recognized as of 7 years postoperatively.

Case 2

A 62-year-old woman with no contributory medical or surgical history presented with the right thigh pain that had gradually worsened over 3 years. No paralysis was apparent in either lower limb. MRI on admission demonstrated a cystic lesion in the conus medullaris spanning from the 12th thoracic to first lumbar level [Figures 3a-c]. Neurological symptoms completely resolved after surgical intervention comprising laminectomy, cyst fenestration, and cyst-subarachnoid shunt placement using the syringo-subarachnoid shunt tube (diameter, 1.2 mm; length, 40 mm; Createmedic Co.). Postoperative MRI after 2 years revealed complete regression of the dilation with no evidence of relapse [Figure 3d]. No recurrence of neurological symptoms has been recognized as of 5 years after surgery.

Case 3

A 67-year-old woman with no contributory surgical or medical history presented with numbness in both lower limbs and weakness in the left lower limb. MRI on admission showed findings similar to those in Cases 1 and 2 [Figure 4]. She was discharged following laminectomy, cyst fenestration and cyst-subarachnoid shunt placement using the syringo-



Figure 1: Results of magnetic resonance imaging (MRI). (a) T2-weighted sagittal image from 12 years before this presentation shows cystic dilatation of the conus medullaris. (b-1) T1-weighted image. (b-2) T1-weighted image with gadolinium enhancement. (b-3) T2-weighted image. (c) Postoperative T2-weighted images obtained 3 years after surgical treatment reveal regression of the dilatation without evidence of relapse (white bold arrow).

subarachnoid shunt tube (diameter, 1.2 mm; length, 40 mm; Createmedic Co.). The postoperative course was uneventful. MRI 1 year after surgery did not show any growth of the cyst lesion [Figure 4]. No recurrence of neurological symptoms has been recognized as of 4 years after surgery.

DISCUSSION

The VT is an ependymal-lined cavity in the conus medullaris that is anatomically continuous with the central canal

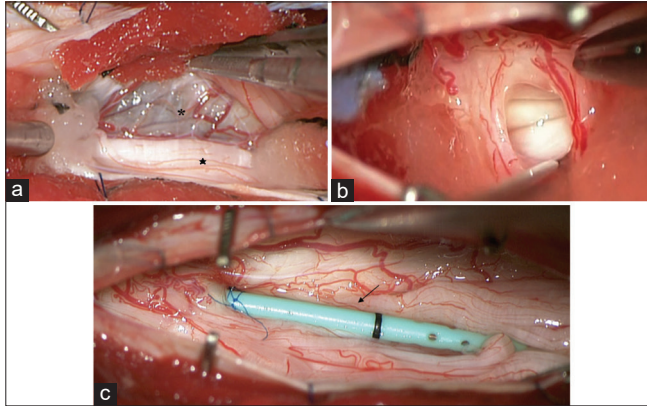


Figure 2: Cystic dilation of the ventriculus terminalis from an intraoperative view. (a and b) On opening the dura, the expanded conus began herniating outward under pressure, which was relieved by fenestration. (c) Cysto-subarachnoid shunt was created using a syringo-subarachnoid shunt tube (diameter: 1.2 mm, length: 40 mm). Black asterisk: Cyst wall, black star: Cauda equina, black arrow: Syringo-subarachnoid shunt tube.

of the spinal cord. This structure usually forms during embryogenesis and subsequently disappears during the regressive differentiation period.^[1-4,8-10] Expansion in adulthood is a very rare but known phenomenon, although the underlying mechanisms are still unclear.^[1,8] Nassar *et al.* reported that dilatation of the cavity can be caused by compression, vascular injury, inflammatory disease or ischemic necrosis.^[5] Sigal *et al.* suggested that physiological changes to cerebrospinal fluid following trauma can lead to abnormal closure and subsequent separation of the cavity, causing the formation of an enlarging VT.^[7] In our cases, none of the three patients had any history or evidence of congenital anomalies such as Chiari malformation or spinal dysraphism, vascular injury, inflammatory disease, trauma, or medical treatments, and none of the patients had undergone surgical interventions for other spinal disorders. In addition, in Case 1, MRI performed for other reasons 12 years before presentation indicated the presence of asymptomatic VT. Although no specific cause was identified, these images confirmed enlargement of the VT to become symptomatic. The causes of VT expansion remain unknown. As in Case 1, the expansion of VT may worsen over time, requiring long-term follow-up even in asymptomatic patients.

Optimal treatment for asymptomatic patients seems to be conservative, but the best treatment strategy for symptomatic enlarging VT has yet to be clarified.^[1-4,8-10] In the previous reports, patients with focal neurological deficits attributable to enlarging VT seem to be best handled surgically. A combination of laminectomy, myelotomy, and subsequent fenestration of the cyst wall is the most widely



Figure 3: Preoperative sagittal and axial spinal magnetic resonance imaging demonstrating a cystic lesion near the conus medullaris. (a) Sagittal T1-weighted image with non-contrast sequence. (b) Sagittal T2-weighted image. (c) Axial T2-weighted image. (d) Postoperative sagittal T2-weighted image 2 years after surgery reveals complete regression of the dilation (white bold arrow).



Figure 4: Sagittal and axial magnetic resonance imaging on admission demonstrating a cystic lesion near the conus medullaris. (a) Sagittal T1-weighted image with non-contrast sequence. (b) Sagittal T2-weighted image. (c) Axial T2-weighted image. (d) Postoperative sagittal T2-weighted image 1 year after surgery shows complete regression of the dilation (white bold arrow).

used treatment.^[3,4] In the previous reports, Pencovich *et al.* showed that a cyst fenestration without cystic-subarachnoid drainage resulted in a complete recovery in 52% of patients and a partial recovery in 43% of patients.^[6] On the other hand, Zhang *et al.* reported that cystic-subarachnoid shunting using a T-catheter was useful for cystic dilation of the VT.^[11] In the present three cases, surgical creation of a cyst-subarachnoid shunt using a syringo-subarachnoid shunt tube proved successful. During a mean follow-up of 64 months, relief from clinical symptoms was observed in all three patients. No patients experienced symptom recurrence during follow-up, and postoperative MRI revealed no lesion regrowth. We therefore consider that as a surgical treatment for symptomatic enlarging VT, cyst-subarachnoid shunt may be minimally invasive and reliable and associated with a lower rate of recurrence.

Several limitations to our report must be kept in mind. At present, symptomatic enlarging VT is a rare neurological condition, and few cases have been reported. Further research, accumulation of more cases, and longer follow-up of patients are required to obtain a better understanding of the pathological conditions and appropriate therapies associated with this entity.

CONCLUSION

We have reported three cases of symptomatic enlarging VT treated with cyst-subarachnoid shunt using a syringo-subarachnoid shunt tube. Symptomatic enlarging VT is an extremely rare cause of conus medullaris syndrome and the

optimal treatment strategy remains unclear. Our successful surgical management of these three cases by cystic-subarachnoid shunting using a syringo-subarachnoid shunt tube for drainage of the enlarging VT provides a potential option for treating symptomatic enlarging VT.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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