Case Report

The cystic dilation of ventriculus terminalis with neurological symptoms: Three case reports and a literature review

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Context: The ventriculus terminalis (VT) is a very small ependymal-lined residual lumen in the conus medullaris. It is normally present in all subjects during fetal development. VT in adults appears as an unusual pathology with an uncertain pathogenesis.

Findings: In this paper, we described three case reports of symptomatic fifth ventricle cystic dilations. All of them were female and their mean age was 59 years. We treated them surgically and all three patients were improved based on clinical and imaging assessments.

Conclusion: Our cases suggested that surgical decompression was a safe and effective treatment in symptomatic patients and the neurosurgeons should be aware of such rare situations. A complete list of differential diagnosis about other cystic dilations of the conus medullaris should be emphasized to select the correct clinical approach.

Keywords: Ventriculus terminalis, Cyst, Spinal cord

Introduction

The ventriculus terminalis (VT), also known as the “fifth ventricle,” is a very small, ependymal-lined residual lumen in the conus medullaris. VT is normally present in all subjects during fetal development,^{1,2} but it appears as a rare pathology in adults^{2,3} with an unclear pathogenesis.³

The asymptomatic localized dilatation of the VT is a normal developmental phenomenon that can be seen using Magnetic Resonance Imaging (MRI).⁴ The physiological role of the VT is uncertain. As the VT is the terminus of Reissner's fiber, a thread-like structure that extends from the epithalamus, it may act as a mechanoreceptor to indicate variations in cerebrospinal fluid (CSF) pressure.⁵

This paper describes three cases of symptomatic fifth-ventricle cystic dilations.

Case reports

Case 1 was a 62-year-old female with low back pain radiating to both limbs. She had been symptom-free

until six months earlier. There were no positive findings after a physical examination of sensory, motor, gait and sphincter functions. An evaluation of her superficial and deep tendon reflexes did not demonstrate any abnormality. The diagnosis was made only after a dorsolumbar MRI (Figure 1), which confirmed the cystic dilation of the VT with an intensity similar to that of CSF. In fact, the intracystic fluid showed the same characteristics as normal CSF: a hypointense T1-weighted sequence, an isointense proton density-weighted sequence, and a hyperintense T2-weighted sequence. After a laminectomy and a cyst fenestration, the patient was discharged without any focal neurological defect.

Case 2 was a 64-year-old female with lower back pain, paresthesia, pain, and a weak lower-left limb for two years. After a radiological evaluation, she was referred for a neurosurgical assessment because of her progressive clinical deterioration. The neurological examination showed gait disturbance, muscular hypotrophy, and lower-left limb hypotonia without any sphincter dysfunction. The patient's dorsolumbar MRI had findings similar to the MRI of Case 1. Although her symptoms improved after a surgical intervention with a

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Figure 1 The cystic dilation of the ventriculus terminalis with an intensity similar to that of CSF. 1A shows a T1 (Case 3), 1B shows a T2 (Case 1), and 1C shows an axial T2 section (Case 1).

laminectomy and cyst fenestration, she had a residual weakness in her lower-left limb.

The last case, Case 3, was a 51-year-old female patient who had lower back pain, neurogenic claudication and sensory symptoms of lower limbs for two past years. Although there was no positive finding on a neurological examination, the MRI (Figure 1a) showed the cystic dilation of the VT with an intensity similar to CSF. She was discharged following a T12 laminectomy and a cyst fenestration.

None of the patients had a history of trauma or evidence of Chiari malformations or spinal dysraphisms, and none of the patients had undergone surgical interventions for spinal problems. In examining their skin in the lumbar, dorsal, and cervical areas, no skin lesions associated with occult spinal dysraphisms, such as a macule, a dermal sinus, fleshy masses, hypertrichosis, hemangioma, or a mass containing soft tissue were observed. Moreover, in the MRIs their filum terminales had no fat infiltrations.

No lesions had been reported in any of the patients in plain radiographic studies done by radiologists except for degenerative changes and the reduced height of some of the lower intervertebral discs. None of the

patients had spina bifida occulta. The lower ends of their conus medullaris were placed normally, and were parallel to the L1–L2 disc spaces. No abnormal enhancements of their cystic walls or their cysts were observed after contrast injections (Figure 2).

The surgical technique consisted of a dorsolumbar (T12 or L1) laminectomy followed by cyst aspiration and a longitudinal (approximately 5 mm) myelotomy (Fig. 3). Each patient's filum had a normal thickness during surgery, and no one's filum was dissected. No intraoperative adhesions or webs in the arachnoid membrane or spinal cord were found. There were no abnormal tissues or nodules within the internal walls of cysts. A clear and uncolored liquid similar to CSF was observed within the cysts. The post-operative course was satisfactory for every patient. We prescribed 100 mg of Gabapentin every eight hours for two months after each surgery; none of the patients developed a new pain that could be attributed to the myelotomy. By the time they were discharged from the hospital, all patients had experienced an immediate clinical improvement. A 28-month follow-up showed that the clinical improvement was sustained except for the weakness in the left limb of the patient in Case 2. MRIs of the patients during

Figure 2 No abnormal enhancement of the cystic wall or the cyst itself can be seen after a contrast injection (Case 3).

Figure 3 The cystic dilation of the ventriculus terminalis from an intraoperative view (Case 2).

Figure 4 MRIs of patients during follow-up periods (two years after their surgeries), which show the significant reduction of their cysts' sizes, from sagittal (A) and axial (B) views.

their follow-up periods confirmed significant reductions in the sizes of cysts (Figure 4).

Results

All patients in the study were symptomatic. Their mean age was 59 years, and all of them were female. None of them had a history of spinal trauma, spinal dysraphisms, or evidence of Chiari malformations, and none of them had undergone surgical treatments for spinal diseases. The patients were diagnosed using MRIs, and their treatments consisted of laminectomies and a dorsal myelotomies. Remarkably, all of patients were ameliorated in terms of clinical and imaging assessments. None of the patients developed complications that could be attributed to their myelotomies. Therefore surgical decompression could be a safe and effective treatment for the symptomatic patients.

Discussion

Syringomyelia should be clearly differentiated from a dilated ventriculus terminalis. The later occurs immediately cephalad to a normally located conus medullaris, is not associated with other dysraphic anomalies and is not related to history of trauma and surgical intervention and is non-progressive on follow-up MRI.⁴ According to Coleman *et al.*⁵ Stilling described a VT in 1859, and in 1875 Krause identified it as a true ventricle lined by ciliated ependymal cells. Krause named it the “fifth ventricle.” The development of the VT, or the

fifth ventricle, can be considered a normal process of embryonal development.⁷ The origin of the VT can be traced to the embryonic development of the spinal cord, which can be subdivided into two stages: neurulation and canalization or retrogressive differentiation.⁸ Neurulation starts when the embryo is three-weeks old, and it is responsible for the formation of the neural tube through the progressive closure of the neural plate and its separation from the overlying ectoderm. The distal cord develops from a caudal cell mass that forms from the neuralepithelium and a notochord caudal to the primary neural tube. The caudal cell mass develops microcysts that coalesce to form an ependyma-lined tube that fuses with the central canal of the neural tube. With differentiation, the caudal end of the cell mass involutes, leaving a focal prominence in the ependyma-lined canal in the conus medullaris with a fibrous tail, which is the filum terminale.⁷ The VT appears after approximately 45 days (between the 43rd and the 48th day) after conception.⁸

The presence of a fifth ventricle has been described in literature as a normal developmental phenomenon, especially in newborns and infants.⁸ The longitudinal diameter of the terminal ventricle is 8–10 mm, and its transverse diameter is 2–4 mm.¹⁰ The VT reaches its final size during the second year of life.¹¹ A pathological study by Choi *et al.*¹² demonstrated that the VT has always been detectable through the histologic examination of the adult or pediatric conus medullaris, while

neuroimaging techniques can barely detect it. In one magnetic resonance series, it was identified in 2.6% of children under five years of age;⁵ but it rarely persists through adulthood.¹³ The persistence of a fifth ventricle in adulthood, especially when unaccompanied by other malformative pathologies of the central nervous system, is extremely rare.⁷ Despite this, the symptomatic dilation of the VT can be seen in young adults.^{13,14} By 2013, 33 cases had been reported,¹³ and after that year, 10 cases had been reported.^{15–18} MRI is the best modality for studying the cord and its syrinx, but in infants whose posterior elements of the spine have not become bony, an ultrasound can also be used to check the status of the terminal ventricle.¹⁹

The mechanism that determines the dilation of the VT is still unclear.² It might be caused by the absence of, or the secondary closure of, communication with the central spinal canal.^{1,3} Nassar *et al.*²⁰ reported that cavitations can be produced by compression, vascular disturbances, inflammatory diseases, or the ischemic necrosis of the spinal cord. Sigal *et al.*¹⁰ postulated that physiological changes in CSF that are a result of trauma can lead to the abnormal closure and subsequent isolation of the VT, precipitating the enlargement of the VT into a cyst. In a postmortem study by Kernohan *et al.*²¹ the VT was reported to be a “true ventricle” that communicated with neither the subarachnoid space nor the central canal of the spinal cord. Based on this fact, and due to site of the cyst within the conus medullaris, the theory of a loss of communication with the central spinal canal may have little significance.

Another interesting theory is venous hypertension. Srivatanakul *et al.*²² found that venous hypertension in the spinal cord induced the development of syringomyelia, which in three cases resolved after embolization. Due to the appearance of normal CSF in the cyst, Liccario *et al.*⁷ referred to a transcystic pressure gradient as a cause of ventricular dilation. According to Celli *et al.*²³ fibrotic and arachnoiditic processes, secondary to post-traumatic meningeal hemorrhages and transcystic pressure gradient modifications caused by post-traumatic stenosis and deformities of the vertebral canal, can cause cyst enlargement.

Another explanation could be the existence of non-communicating accessory canals in the conus medullaris that undergo secondary dilatation,¹⁰ such accessory canals have been discovered in 35% of 4–32 mm embryos.²⁴ This theory can explain both asymmetric and paramedian cysts.

Ventricular dilatation appears on MRIs as a small, roundish cavity with regular borders, which has no septum. The cavity is filled with a liquid of the same intensity as CSF in both T1 and T2 sequences without

the enhancement of a paramagnetic contrast medium after an injection.⁶ During differential diagnoses, localized syringohydromyelia and cystic tumors should be considered in the conus area. The isolated localized lumbosacral syringohydromyelia of the spinal cord is very rare, accounting for 2.5% of all such lesions. Almost all of those lesions are associated with evidence of a dysraphism, a pes cavus, or a Chiari I malformation.²⁵ However, gadolinium enhancements, higher-intensity fluid contents of cysts, and irregular margins with a soft tissue compartment are evidence of tumors associated with the cystic dilation of the spinal cord.²⁶

Various symptoms and signs described in patients included low back pain, sciatica,^{10,23} progressive scoliosis, limb paresis and pain, and sphincter dysfunction.⁵ According to Suh *et al.*² clinical symptoms can correlate with the size of the conus lesion; the larger the size of the conus lesion, the greater the severity of the clinical symptoms. However, Ganau *et al.*³ found that cyst size did not appear to be directly proportional to the clinical complaint but was instead directly proportional to the clinical outcome, given that the patients with less satisfactory results were the patients with the larger cysts. The therapeutic approach was selected based on clinical manifestations and image findings.

Batista *et al.*²⁷ divided patients into three groups. Type I was patients that show non-specific neurological symptoms or have non-specific complaints, including sciatica, low back pain, and inferior limb pain. Type II was patients that have paresis, muscular atrophy, a focal neurological deficit, altered deep reflexes, or sensory disturbances. Type III was patients that have sphincter dysfunctions, such as bowel and/or bladder incontinence. Patients exhibiting Type I symptoms can be controlled without surgery, but the other groups should be treated with a surgical intervention.

Ganau *et al.*³ further divided Type I symptoms into two subgroups. Type Ia was patients that have stable clinical symptoms within a couple of months of observation, and they have no clear relation to the VT due to the coexistence of other spinal disorders. Type Ib was patients that have non-specific but worsening symptoms and require a surgical evacuation of the cyst. As cystic dilation of the ventriculus terminalis (CDVT) is one of the very rare causes of conus medullaris syndrome and the development of neurological symptoms, its therapeutic strategy is unclear.¹⁷ However, given the fact that most reported cases are single, determining the outcome of the treatment and comparing different treatments is difficult.

A laminectomy, a myelotomy, and the subsequent fenestration of the cyst wall is the most widely used

treatment for patients.^{4,18,27,28} This method is associated with good outcomes for those with motor and sphincter disorders, although its effect on pain is controversial.³ No recurrences have been reported during patient follow ups.²⁷ In one study, a cyst fenestration resulted in a complete recovery in 52% of patients and a partial recovery in 43% of patients.¹³ Batista *et al.*²⁷ performed a hemilaminectomy and a dorsolateral myelotomy on two patients, which produced favorable results. A shunt placement was used in some cases for cyst drainage.²⁹ The shunt placement was associated with a high rate of complications and failure in long-term follow-ups.^{30,31} Zhang *et al.*¹⁸ performed a laminoplasty and a cyst fenestration on six patients, after which a T-catheter was inserted inside their cysts; the procedures were associated with favorable outcomes after a 41-month follow-up. The use of intraoperative neuromonitoring, such as electromyography, motor evoked potential, and somatosensory evoked potential will significantly reduce the risk of post-operative neurological complications.³²

In recent years, some authors have conducted the percutaneous aspiration of cysts using real-time MRI. The aspirations were associated with favorable primary outcomes.³³

The authors of this paper agree with Agrillo's,¹¹ and Sigal's,⁹ claim that due to characteristic MRI appearances of the lesions, there is no need for biopsies. Therefore definite diagnosis can be achieved without the need to perform biopsies.

Conclusion

The cystic dilation of the VT is a rare disease with an unclear pathogenesis. This review of three cases with symptomatic fifth-ventricle cystic dilations shows that the cysts are benign lesions. None of the patients had a history of trauma, spinal dysraphisms, or evidence of Chiari malformations. None of them had undergone surgical intervention for spinal problems.

This study suggests that surgical decompression is a safe and effective treatment for symptomatic patients. The importance of an exhaustive differential diagnosis considering other cystic dilations of the conus medullaris should be emphasized for a correct clinical approach to this type of disease. Neurosurgeons should be aware of such rare situations.

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