CYSTIC DILATION OF THE CONUS VENTRICULUS TERMINALIS PRESENTING AS AN ACUTE CAUDA EQUINA SYNDROME RELIEVED BY DECOMPRESSION AND CYST DRAINAGE: CASE REPORT

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Received, June 27, 2005.
Accepted, October 27, 2005.

OBJECTIVE AND IMPORTANCE: The ventriculus terminalis of the conus, or "fifth ventricle" refers to the ependymal-lined space in the middle of the conus that is present in childhood and whose persistence into adulthood is rare. A number of cases of cystic dilatation of the ventriculus terminalis have been described in adulthood. Patients tend to present with either pain alone or gradually progressive conus or cauda equina syndromes with varying degrees of recovery after cyst drainage. Presentation with an acute cauda equina syndrome and its successful surgical management has not been previously reported.

CLINICAL PRESENTATION: A 57-year-old woman experienced back pain and bilateral sciatica ascribed to diabetic neuropathy for 2 years. Over a 24-hour period she developed bilateral lower extremity weakness, saddle anesthesia, and bowel and bladder incontinence. Lumbosacral magnetic resonance imaging demonstrated a large cystic dilatation of the ventriculus terminalis.

INTERVENTION: She was taken for emergency surgical decompression and cyst drainage. Immediately after surgery, she experienced significant increase in lower extremity strength and has since regained continence.

CONCLUSION: Cystic dilation of the ventriculus terminalis should be part of the differential diagnosis for a cauda equina syndrome; surgical decompression with simple cyst drainage can result in excellent clinical results.

KEY WORDS: Cauda equina syndrome, Conus, Cyst, Fifth ventricle, Surgery, Ventriculus terminalis

Neurosurgery 58:585-586, 2006 DOI: 10.1227/01.neu.0000197486.65781.88 www.neurosurgery-online.com

First described by Stilling (1, 3) in 1859, the ventriculus terminalis is an ependymal lined cavity in the conus medullaris that becomes identifiable between Days 43 and 48 after conception as part of the caudal neural tube. Present during fetal development, this space typically regresses after birth, although it has been detected in magnetic resonance imaging (MRI) scans in 2.6% of children under the age of 5 years imaged for unrelated symptoms. Its detection is extremely rare in adults, with 18 cases reported to date (5). Krause identified the microscopic pathology of the ventriculus terminalis as a ventricular structure lined by ciliated ependymal cells, and termed it the "fifth ventricle" (3). Whether or not cystic dilatation of the ventriculus terminalis (CDVT) simply represents syringomyelia of the conus medullaris is controversial and relates to the pathogenesis of the condition, which remains unknown.

CDVT represents a failure of regression of the ventriculus terminalis; possible etiologies include failed fusion between the cranial and caudal neural tubes secondary to congenital disorders, trauma, and ischemia (2). An association between CDVT in children and the full spectrum of spinal dysraphism and tethered cords has been identified, but the exact causation has not been identified. Because descriptions of CDVT in adults has been limited to case reports and small series, true associations are harder to identify. In one literature review of 11 reported cases of CDVT in adulthood,

however, two patients harbored sacral lipomas and one had a Type I Chiari malformation (1).

CDVT in infancy and childhood, not associated with spinal dysraphism, is most often an incidental finding and conservative nonoperative management has been advocated. Of the published cases of CDVT in adults, some have been incidental findings or presenting with pain only, whereas others have presented with insidiously progressive neurological deficits attributable to compression of the conus and cauda equina. We describe the first case presenting with an acute cauda equina syndrome and treated successfully with surgery.

CASE REPORT

A 57-year-old woman with a history of diabetes, hypertension and hypercholesterolemia presented to the emergency room complaining of a recent episode of bowel and bladder incontinence and rapidly progressive lower extremity weakness. She also noted buttock, thigh, and perianal numbness. For the past 3 to 4 years, the patient had been experiencing lumbar and bilateral sciatic pain as well as increasing difficulty with her gait that was ascribed to diabetic neuropathy. She denied history of antecedent trauma.

On examination, the patient was unable to lift her lower extremities against gravity. Distal lower extremity power was 4+/5. Sensation was grossly normal, although no deep tendon reflexes of the lower extremities could be detected; her toes were downgoing. She had no physical stigmata to suggest occult spinal dysraphism.

Lumbosacral MRI scans with and without gadolinium demonstrated a large intramedullary cyst of the conus without evidence for contrast enhancement (*Fig. 1A*).

The cyst expanded the conus and its fluid content was similar to cerebrospinal fluid (CSF) on all sequences (*Figs. 1, A–D*). There was no evidence for spinal lipoma or dysraphism. Craniocervical imaging failed to reveal evidence for a Chiari malformation or other intracranial abnormality. The patient was taken to the operating room the following day, where she underwent a T11–L1 bilateral laminectomy. Upon opening the dura, the expanded conus began herniating outward under pressure, which was relieved by a midline dorsal myelotomy. CSF-colored fluid was drained and the myelotomy extended. Formal marsupialization was not performed. No abnormal tissue suggestive of tumor was identified. Pathological analysis of a small piece of the cyst wall revealed flattened epithelium-lined ependymal cells situated within the spinal cord parenchyma (*Fig. 2, A* and *B*), thus differentiating this from a more traditional syrinx in which the epithelium is in continuity with the central canal, or a simple arachnoid cyst.

Postoperatively, the patient experienced marked improvement in her lower extremity strength and has enjoyed subjective increased sensation of her buttocks and lower extremities. At a 3-month follow-up examination, she had no further episodes of urinary or bowel incontinence, and was able to ambulate with the assistance of a walker. Her lower extremity sensation is grossly normal and she has improved temperature and light touch sensation of the perineum. Immediate postoperative MRI scans revealed collapse of the previously identified cyst (*Fig. 3A*). Follow-up MRI scans 1 month later revealed minimal reaccumulation of fluid within the cyst (*Fig. 3B*).

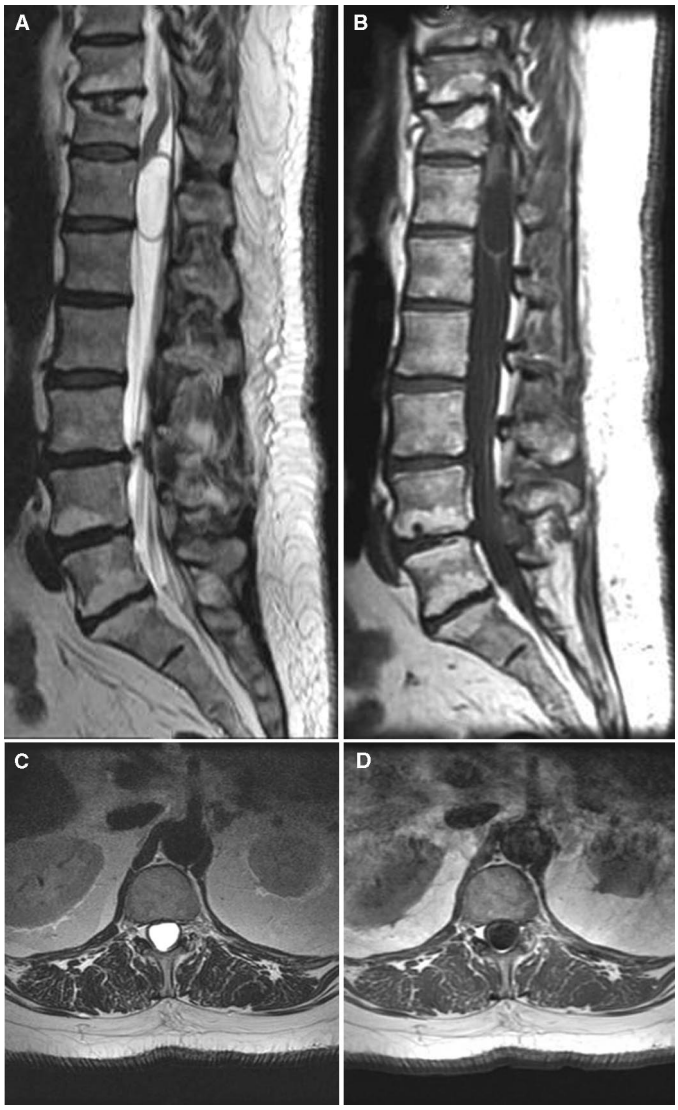

FIGURE 1. *Sagittal T2-* (A) *and T1-weighted* (B) *MRI images displaying a cystic cavity in the region of the conus medullaris. Although the T2-weighted image suggests an intramedullary lesion pushing the conus forward, the T1-weighted image shows the filum terminale emanating from the center of the caudal portion of the cyst, more suggestive of a CDVT. Axial T2-weighted* (C) *and T1-weighted* (D) *MRI images confirm the intra-axial cyst at the level of the conus. Notice that the cyst fluid is similar to that of cerebrospinal fluid on all sequences.*

DISCUSSION

CDVT presenting in adulthood is rare, with eighteen previous cases described in the literature (1, 2, 4–9). Half of these

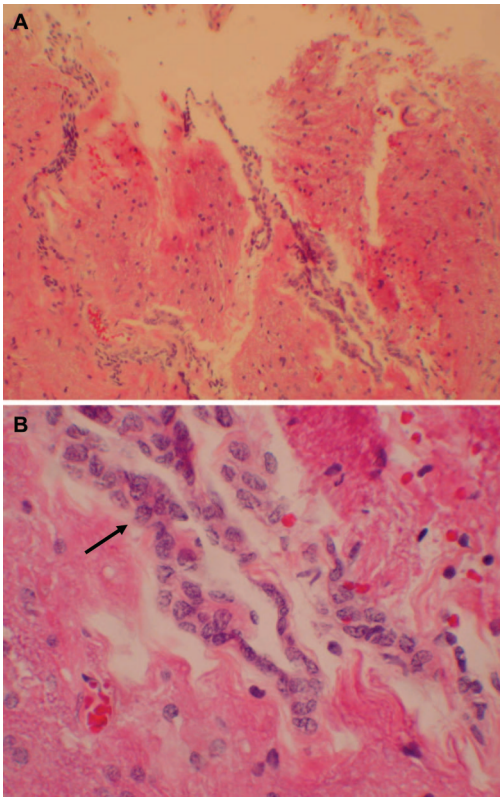

FIGURE 2. *Low (*A*) and high (*B*) magnification hematoxylin and eosin stained sections of resected tissue from the conus demonstrates a layer of flattened ependymal epithelium (*B, arrow*) surrounded by normal white matter (original low magnification, ×25, original high magnification, ×100).*

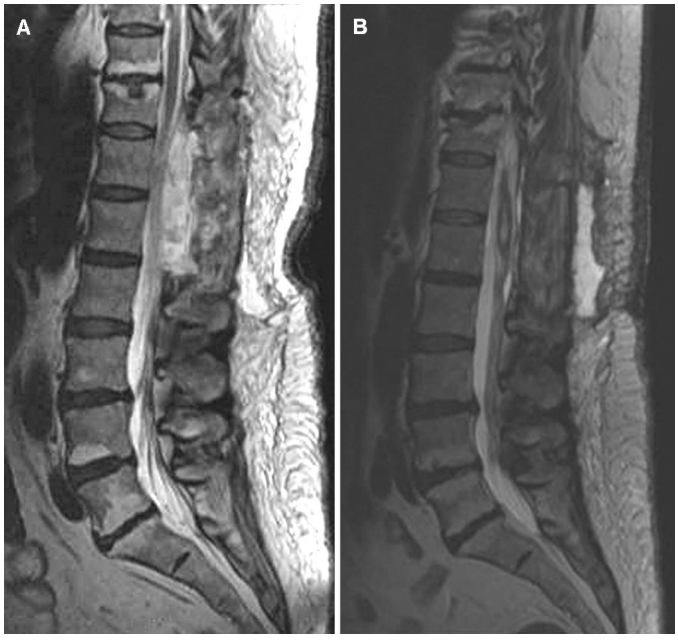

FIGURE 3. *Postoperative T2-weighted sagittal images obtained immediately (*A*) and 1 month (*B*) after surgery showing successful bony decompression and collapse of the cyst. The conus resumed its normal midline location within the spinal canal. A small amount of cystic fluid reaccumulation is seen at 1 month.*

reported cases have been published in the radiological literature, where the sonographic and MRI appearance has attracted considerable interest (2, 6, 10). Several reported cases of CDVT have been asymptomatic or accompanied only by lumbar pain and sciatica. Some authors, particularly in the radiological literature, have described managing such patients conservatively and, after documenting lack of clinical and radiological progression, have argued strongly against surgical treatment for this disorder (1, 5, 6, 8). Other instances of adult-onset CDVT have displayed the full spectrum of neurological deficits related to conus or cauda equina malfunction. These have typically undergone surgical decompression with cyst marsupialization, usually with good results (7, 9).

Presentation with urinary sphincter disturbance is not uncommon, as compression from pressure within the cyst causes injury to the conus itself and/or the surrounding nerve roots. Typically this finding is either mild and not associated with other symptoms and signs suggestive of a more extensive conus or cauda equina syndrome, or is simply very gradually progressive and, therefore, does not cause alarm (8). Previous presentation with an acute cauda equina syndrome has not been recorded. The mechanism whereby an indolent CDVT becomes acutely symptomatic, as in our case, is not known and presumably relates to the mechanism of formation and growth. Hypotheses that have been put forth hinge on whether CDVT is a hydromelic process, with expansion of the central canal, or a syringomyelic one, involving the spinal parenchyma separate from the central canal. Current opinion favors a hydromelic process in which the communication between the ventriculus terminalis and the central canal is blocked secondary to dysembryogenetic failed regression (5, 8). Some, but diminished, communication between the cyst and the central canal exists during the life of the individual, explaining the MRI scans and operative findings of CSF-like fluid within the cyst. If, for some reason, communication between the CDVT and the central canal is further blocked, a transcystic pressure gradient may develop leading to new symptoms. Interestingly, no documented radiographic expansion of the cyst has been reported.

The first surgical treatment of this entity was described in 1968, in a report documenting treatment of two such cases, both presenting with sciatica and gradually progressive lower extremity neurological dysfunction. In both cases, myelography diagnosed an intramedullary lesion of the conus and surgery consisting of simple cyst drainage resulted in good neurological recovery (7). This article furnished the first operative photos of CDVT. Since that time, at least 12 other patients with neurological dysfunction and CDVT were operated upon, with excellent results achieved in 12 out of 14 cases (5).

CDVT Represents an Additional Entity that May Cause Cauda Equina Syndrome

We suspect that an acute cauda equina syndrome resulting from CDVT behaves similarly to that from other etiologies and

that urgent decompression should be undertaken to avoid catastrophic and permanent symptoms from cauda equina injury. Furthermore, this case suggests that all symptomatic CDVTs should be drained, given the unpredictable nature of the condition and the relatively low risks of the surgical procedure. Simple wide laminectomy, midline myelotomy and cyst drainage has proven adequate, although some have used marsupialization or cysto-subarachnoid tube drainage adjunctively. As with other central nervous system cysts, long-term follow-up will be important to monitor for recurrence.

This case also emphasizes the importance of obtaining imaging studies for anyone with persistent complaints of lumbar pain, sciatica, or lower extremity neurological dysfunction, regardless of possible additional metabolic etiologies. The MRI appearance of an intramedullary conus cyst containing fluid that follows CSF patterns and no enhancement with gadolinium is pathognomonic for CDVT.

CONCLUSION

CDVT is a rare disorder in adulthood, representing a failure of embryologic regression during spinal cord formation. Although the isolated finding of CDVT is usually benign in children, in adults the clinical presentation and progression is unpredictable. We describe a case presenting with an acute cauda equina syndrome. The MRI appearance of this lesion is pathognomonic and neurosurgeons should be aware of this entity as surgical decompression with simple cyst drainage is both safe and effective.

REFERENCES

1. Celli P, D'Andrea G, Trillo G, Roperto R, Acqui M, Ferrante L: Cyst of the medullary conus: Malformative persistence of terminal ventricle or compressive dilatation? **Neurosurg Rev** 25:103–106, 2002.
2. Dullerud R, Server A, Berg-Johnsen J: MR imaging of ventriculus terminalis of the conus medullaris. A report of two operated patients and a review of the literature. **Acta Radiol** 44:444–446, 2003.
3. Kernohan JW: The ventriculus terminalis: Its growth and development. **J Comp Neurol** 38:10–125, 1924.
4. Korosue K, Shibasaki H, Kuroiwa Y, Machi T, Sawada K, Kitamura K, Ikeda J: Cyst of the conus medullaris manifesting amyotrophic lateral sclerosis syndrome. **Folia Psychiatr Neurol Jpn** 35:507–510, 1981.
5. Liccardo G, Ruggeri F, Cerchio LD, Floris R, Lunardi P: Fifth ventricle: An unusual cystic lesion of the conus medullaris. **Spinal Cord** 43:381–384, 2005.
6. Matsubayashi R, Uchino A, Kato A, Kudo S, Sakai S, Murata S: Cystic dilatation of ventriculus terminalis in adults: MRI. **Neuroradiology** 40:45–47, 1998.
7. Nassar SI, Correll JW, Housepian EM: Intramedullary cystic lesions of the conus medullaris. **J Neurol Neurosurg Psychiatry** 31:106–109, 1968.
8. Sigal R, Denys A, Halimi P, Shapeero L, Doyon D, Boudghene F: Ventriculus terminalis of the conus medullaris: MR imaging in four patients with congenital dilatation. **AJNR Am J Neuroradiol** 12:733–737, 1991.
9. Stewart DH Jr, King RB, Lourie H: Surgical drainage of cyst of the conus medullaris. Report of three cases. **J Neurosurg** 33:106–110, 1970.
10. Truong BC, Shaw DW, Winters WD: Dilation of the ventriculus terminalis: Sonographic findings. **J Ultrasound Med** 17:713–715, 1998.

COMMENTS

Brisman et al. have presented a rare clinical case. Cystic dilation of the conus ventriculus terminalis is akin, in a way, to a loculated central canal or a true hydromyelia with lack of communication to the ventricular system rostrally. It is probable that communication to the ventricular system is occluded, hence the entrapped appearance and nature of the lesion. This case serves as an example of yet another variation in the wide spectrum of pathologies categorized under the heading of syringomyelia. The authors are to be congratulated for their presentation of this case.

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Many causes for cauda equina syndrome have been proposed. This case report adds another possibility for the cause of cauda equina syndrome. It is obviously speculative, however, why this dilatation of the ventriculus terminalis presented suddenly. Nevertheless, the authors proceeded with immediate decompression with good results. Neurosurgeons should be aware that, besides the routine causes for cauda equina syndrome, rarely, cystic dilatation of the ventriculus terminalis can also present as cauda equina syndrome.

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The authors report a single case of a patient with a relatively acute onset of significant neurological symptoms referable to a large intramedullary cyst of the conus. The patient was treated with drainage of this cyst and improved markedly. Unfortunately, the clinical follow-up is only 3 months in duration and the follow-up magnetic resonance imaging scan was obtained 1 month after surgery. This report underscores the fact that an intramedullary cyst may be responsible for acute neurological decline. It is curious that the patient experienced a rapid progression of neurological symptoms because one may surmise that the cyst had been present for years and was slowly enlarging. It is difficult to obtain satisfactory long-term clinical outcomes with simple cyst drainage. The follow-up for this patient is very short and, at this point, it is unknown whether this intervention will produce lasting relief.

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Brisman et al. describe a patient who presented with an acute cauda equine syndrome and was found to have a cystic dilatation of the ventriculus terminalis. The patient was treated with an expeditious drainage of the cyst and experienced significant neurological recovery. Although these cysts are usually asymptomatic developmental abnormalities, this case report illustrates the fact that symptoms can develop in adulthood. When symptoms do occur, they may respond favorably to surgical intervention.

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