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Ventriculus Terminalis of the Conus Medullaris: MR Imaging in Four Patients with Congenital Dilatation

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MR findings in four patients with MR evidence of congenital cystic dilatation of the ventriculus terminalis were reviewed retrospectively. The ventriculus terminalis is a small cavity of the conus medullaris that forms during embryonic development as a result of canalization and retrogressive differentiation. The dilated ventriculus terminalis appears on MR images as a small ovoid cavity with regular margination; intraserial fluid resembles cerebrospinal fluid on all MR sequences. After injection of contrast material, MR imaging shows no enhancement of the cyst or its wall, and thus differentiates congenital dilatation of the ventriculus terminalis from cystic tumors, which occur more frequently in this region.

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The ventriculus terminalis is a small ependymal-lined cavity of the conus medullaris [1] that was initially described by Stilling in 1859 [2]. This cavity is only visible on pathologic examination and is not depicted by imaging techniques. We describe the findings in four patients in whom MR imaging revealed a cystic lesion of the conus medullaris that corresponded to congenital dilatation of the ventriculus terminalis.

Materials and Methods

The findings in four patients who showed evidence of dilatation of the ventriculus terminalis on MR images were evaluated retrospectively. All four patients were women, with ages ranging from 35 to 62 years old. All presented with nonspecific neurologic symptoms, including recurrent low back pain, sciatica, and bladder dysfunction. There was no previous history of spine trauma. All patients had conventional radiographs and spinal CT. One patient had myelography. MR imaging was performed in two cases on a 1.5-T unit (Signa, General Electric, Milwaukee) and in two cases on a 0.5-T system (MR Max, General Electric, Milwaukee). Sagittal and axial sequences were obtained with spin-echo T1-weighted sequences, 600/20 (TR/TE), and proton density T2-weighted sequences (2000/20, 90). Post-contrast examinations were performed in all cases. The entire spinal cord was imaged, including the craniocervical junction. Two patients had two MR scans before surgery and a subsequent MR control scan. One of them initially had surgery at the craniocervical junction, but a postoperative MR control scan did not show any modification of the cyst. A second intervention was then performed on the cyst itself in the thoracolumbar region. The other patient had surgery only at the T11-L1 level. In both cases, a large cyst filled with colorless, watery fluid was found in the conus medullaris. Careful inspection of the lesion and intraoperative sonography confirmed the absence of tumor. No biopsy was done of the cyst wall. The cyst was drained into the subarachnoid space. In the other two patients, clinical and MR follow-up findings suggested the diagnosis, and surgery was not needed.

Results

Conventional radiographs were normal except for one patient who had a limited mass effect on the vertebrae at the T12-L1 level. Myelography demonstrated a

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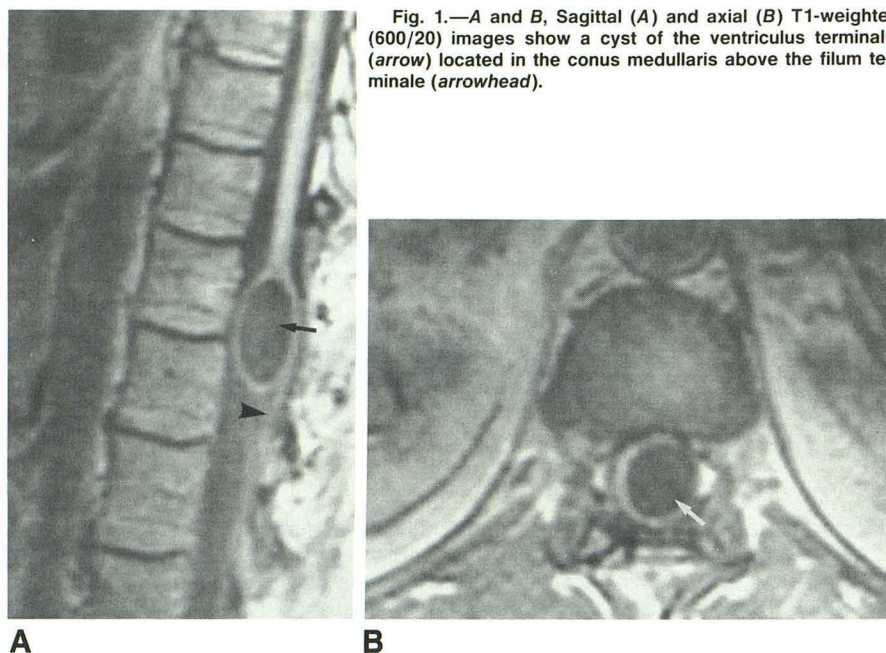


Fig. 1.—A and B, Sagittal (A) and axial (B) T1-weighted (600/20) images show a cyst of the ventriculus terminalis (arrow) located in the conus medullaris above the filum terminale (arrowhead).

large mass of the conus with complete blockage of the flow of contrast material at T12–L1. CT was inconclusive in all cases. In two patients, because of clinical symptoms, CT was performed only in the lower lumbar region. In the other cases, CT showed generalized enlargement of the conus but did not demonstrate definite tumor.

MR findings were similar in all patients (Figs. 1–4), and showed a cystic cavity of the conus medullaris with a craniocaudal dimension of 25–40 mm and a transverse diameter of 17–25 mm. The neural tissue was reduced to a layer less than 2 mm in thickness, because of the mass effect of the cyst. The lower extent of the cavity was the termination of the conus medullaris just above the origin of the filum terminale and nerve roots. The cavity was ovoid with regular margination and with no internal septa. In one case, the cyst presented with a pointed superior extension (Fig. 4). Signal intensity of the intracystic fluid closely resembled that of CSF on all sequences, with low signal intensity on T1-weighted images and high signal intensity on proton density and T2-weighted images. After injection of contrast material there was no abnormal enhancement, either in the vicinity or in the wall of the cyst, thus differentiating this congenital cyst from a cystic tumor. In all follow-up scans, there was no change in the shape or signal intensity of the cyst. The spinal cord was normal in three patients. In the other one, MR disclosed an associated Chiari I malformation.

Discussion

The ventriculus terminalis is formed during fetal life. The embryonic development of the spinal cord consists of two stages called neurulation and canalization/retrogressive differentiation (Fig. 5) [3–5]. Neurulation starts when the embryo is about 3 weeks old (3-mm stage), and corresponds to the

progressive closure of the neural plate into the neural tube, with separation from the overlying ectoderm. The most caudal end of the neural tube forms the posterior neuropore, and closes when the embryo is 4 weeks old (6-mm stage), at the level of the L1 metamere. During the following stage, called vacuolization (Fig. 5A), the neural epithelium, located caudally in respect to the posterior neuropore, fuses with the notochord and forms a caudal cell mass. Then, during canalization, multiple microcysts and clumps of cells form in this mass and coalesce to form an ependymal-lined tube that fuses with the central canal of the neural tube (Fig. 5B). The final stage, called retrogressive differentiation, begins at 5½ weeks: the upper part of the ependymal-lined tube, at level somite 32, dilates (Fig. 5C) and becomes the ventriculus terminalis within the conus terminalis. The most distal caudal cell mass and central lumen undergo a necrosis process that results in the formation of a fibrous structure, the filum terminale (Fig. 5D) into which the ventriculus terminalis may extend. Thus, the spinal cord is formed by a connection between an upper part with a central lumen, the ependymal canal, and a lower part, the conus medullaris, that contains the ventriculus terminalis.

The mechanism that accounts for isolated dilatation of the ventriculus terminalis is still unclear, and to our knowledge has not been explained. Various theories have been proposed to elucidate the pathophysiology of hydromyelia and syringomyelia. Hydromyelia is defined as a dilatation of the ependymal canal, as opposed to syringomyelia, which corresponds to a cystic cavity developing outside the ependymal canal. The theories of Garner [6, 7] and Williams [8] describe a persistent communication between the fourth ventricle and the ependymal canal, with a descending extension of the hydromyelia from the craniocervical junction. They do not, however, explain the location of the cyst in the conus terminalis. Aboulker's theory [9] is based on an obstruction at the craniocervical level (i.e., Chiari malformation). This obstruction

Fig. 2.—A and B, Sagittal T1-weighted (600/20) (A) and T2-weighted (2000/100) (B) images show a thin-walled cyst (arrow) located at T11–T12 level. Its signal intensity resembles that of CSF. The cyst walls (arrowheads) are reduced to a thin layer that measures less than a millimeter.

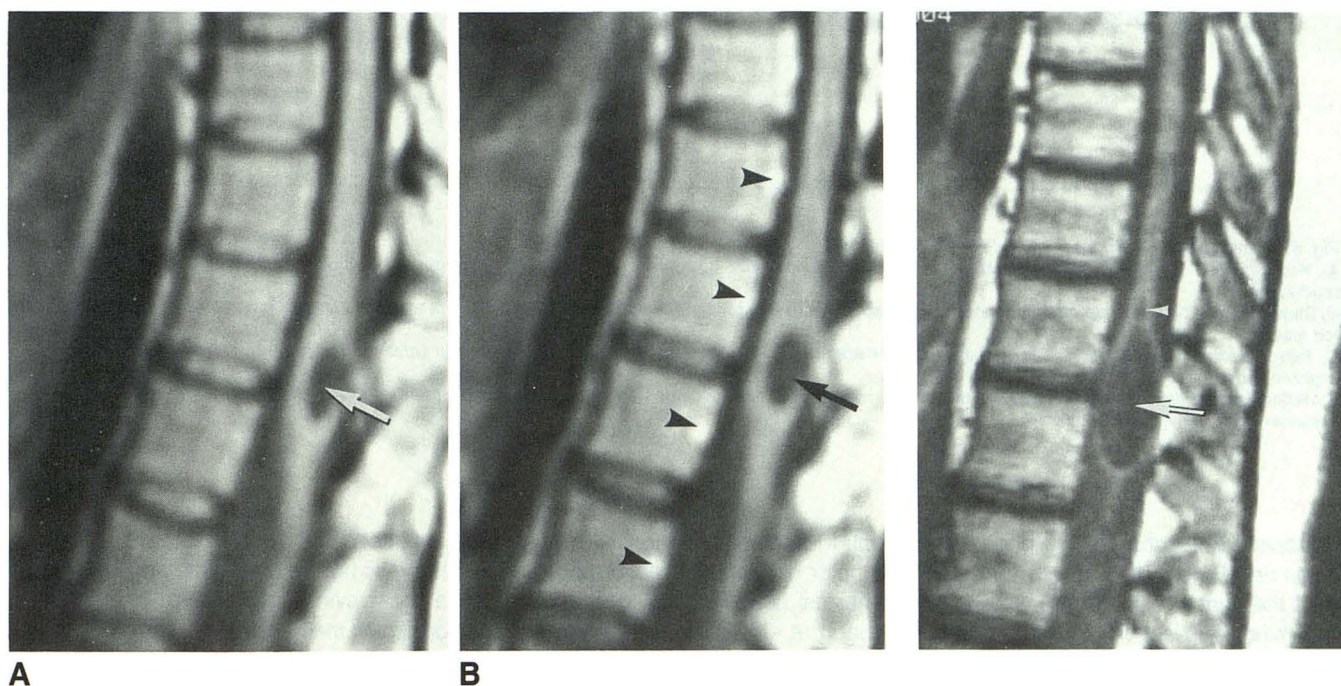
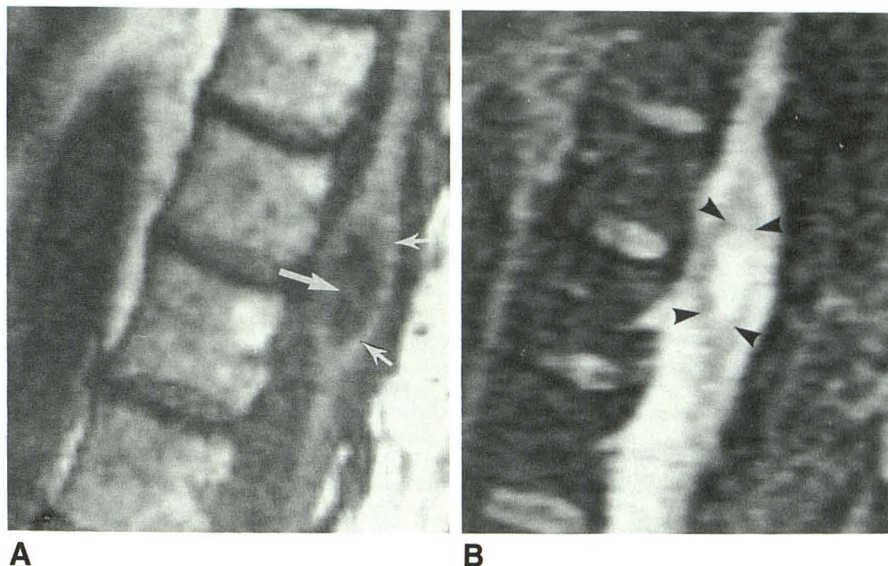


Fig. 3.—A and B, Sagittal T1-weighted images show a cyst of the ventriculus terminalis (arrow) before (A) and after (B) injection of gadopentetate dimeglumine. There is no contrast enhancement of the cyst, but it is seen in the basivertebral veins (arrowheads).

Fig. 4.—Sagittal T1-weighted (600/20) image shows a cyst of the ventriculus terminalis that is ovoid (arrow) with a superior pointed expansion (arrowhead). Surgical findings confirmed the diagnosis.

generates a pressure gradient between the cranial and the medullary subarachnoid spaces with the result that CSF passes into the neural tissue but not into the ependymal canal. This theory better explains the location of the cyst in the conus medullaris, but it cannot account for the fact that the cyst is only located in the ventriculus terminalis. Moreover, one of our patients presenting with an associated Chiari I malformation was operated on at the foramen magnum level. According to Aboulker's theory, surgery should have resulted

in a reduction in the cyst's size. Subsequent MR control scans, however, did not demonstrate any reduction in size of the ventriculus terminalis cyst.

One could speculate that isolated dilatation of the ventriculus terminalis is due to an absence of communication between the ependymal canal of the upper spinal cord and the central canal of the lower spinal cord. Abnormal closure of the ventriculus terminalis could be either congenital or the result of trauma or ischemia. This could explain the absence

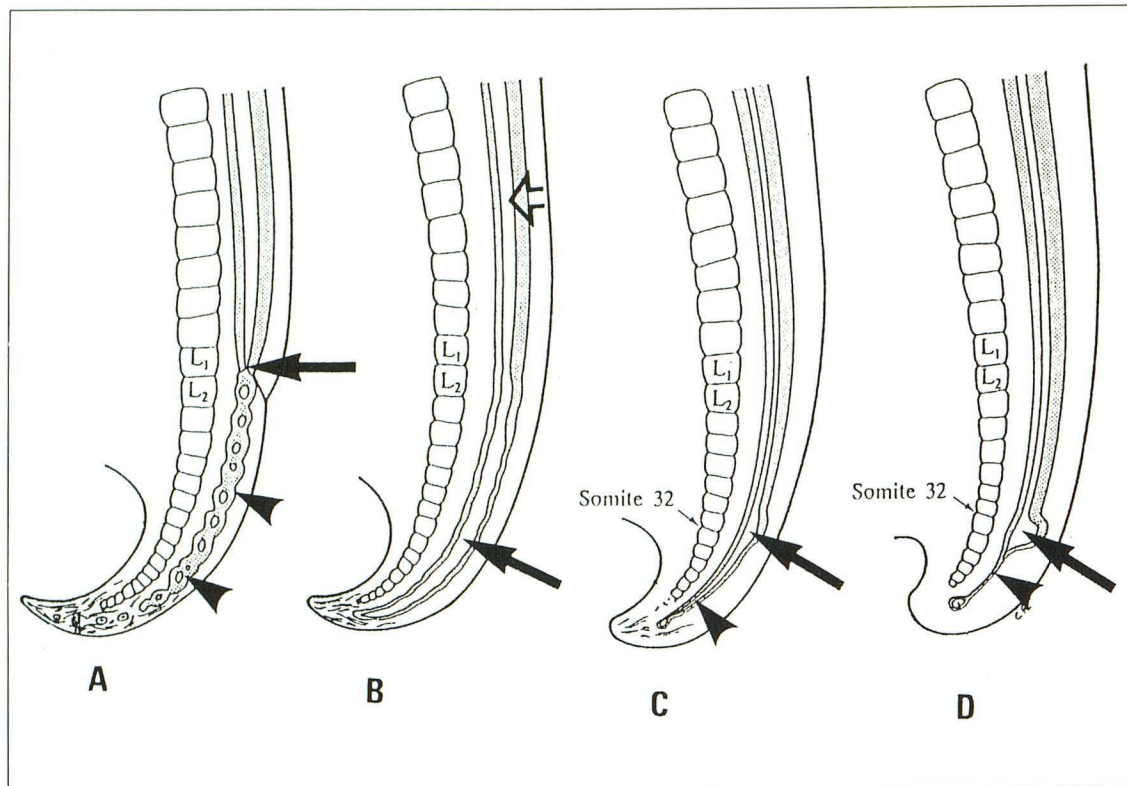


Fig. 5.—Embryonic development of ventriculus terminalis.

A, Vacuolization. This process starts after closure of the posterior neuropore (arrow) and corresponds to the formation of a caudal cell mass (arrowheads).

B, Canalization. The vacuoles coalesce and form an endymal-lined canal (closed arrow) that connects with the central canal of the upper neural tube (open arrow).

C, Retrogressive differentiation. The canal dilates at somite 32 to become the ventriculus terminalis (arrow), and the caudal cell mass begins its regression (arrowhead).

D, Definitive formation of the ventriculus terminalis (arrow) with complete regression of the caudal cell mass that forms the filum terminale (arrowhead). (Adapted from [4] with permission)

of associated dilatation of the endymal canal. In addition, Lendon and Emery [10] have mentioned that major duplication of the central canal is present in 10% of normal adults, and a minor duplication appears in 31%. Raybaud et al. [4] observed accessory canals in 35% of 4- to 32-mm embryos. Thus, another explanation could be the existence of a non-communicating accessory canal in the conus medullaris that undergoes secondary dilatation.

In fetuses and newborns, the conus medullaris may be visualized with sonography [11]. The endymal canal is displayed as an echogenic linear structure, and conceivably could demonstrate congenital isolated dilatation of the ventriculus terminalis. Because clinical symptoms are nonspecific, CT may focus on the incorrect level, missing the conus medullaris, as in two of our patients.

MR imaging localizes the lesion precisely. The fact that the cyst remains confined to the conus medullaris, as shown on follow-up MR scans, makes the diagnosis of syringomyelia unlikely because a syrinx in the lower spinal cord almost always extends superiorly [12]. A congenital origin of the lesion may be suggested when there is an associated con-

genital malformation, as in one of our patients with a Chiari I malformation. The true cystic nature of this lesion is important to recognize because most cystic conus medullaris lesions are tumors and not congenital malformations. The most common tumors are ependymomas (located in the conus medullaris in 50% of cases), followed by astrocytomas and oligodendrogliomas [13]. Intramedullary neuromas are rare [14] as are hemangioblastomas [15]. In conus medullaris cystic tumors, MR imaging would be expected to show contrast enhancement of the solid portion of the tumors, even when they are small, as in hemangioblastomas. In contrast, the cyst of the ventriculus terminalis does not enhance after contrast injection, and the signal intensity mimics that of CSF on both T1- and T2-weighted sequences.

A definitive diagnosis of ventriculus terminalis cyst can only be made on the basis of histopathologic results. Nassar et al. [16] report one patient in whom the microscopic examination of the cyst showed that the wall of the cyst was formed of a layer of endymal cells. Such histopathologic correlation is no longer mandatory, since MR imaging gives sufficient confidence in the diagnosis of ventriculus terminalis cyst. The

therapeutic management of this lesion is not clearly established. Before the advent of MR imaging, conus terminalis enlargement was always a reason to suspect tumor, and surgery was performed. MR imaging can diagnose benign cystic dilatation of the ventriculus terminalis and exclude a tumor. Therapeutic management of cyst of the ventriculus terminalis can thus be based on clinical symptoms and clinical and MR imaging evaluation.

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