

Surgery or not? A case of ventriculus terminalis in an adult patient

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Abstract: The ventriculus terminalis (VT) is an intramedullary cavity containing cerebrospinal fluid (CSF), usually located at the conus, and represents an unsuccessful regression of a normal embryological phase of the development of the neural tube. Symptoms are not univocal and may vary from aspecific signs to sphincter dysfunctions and focal neurological deficits. The correct management of this condition is still an object of debate due to its rarity and its unpredictable behaviour in adult patients. Two different studies have proposed a distinction for patients with cystic dilatation of VT based on their symptoms, suggesting that only those patients with neurological deficits clearly related to the terminal ventricle could benefit from surgery. We describe a case of an adult patient with a progressive dilatation of a diagnosed VT that we treated surgically. A management flowchart for this condition is also proposed based on our experience and a review of the literature.

Keywords: Fifth ventricle; ventriculus terminalis (VT); lumbar spine; conus medullaris

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Introduction

The ventriculus terminalis (VT) is an ependyma-lined space located in the conus medullaris, which represents a normal developmental phase of the neural tube. It appears between the 43th and the 48th day of embryogenesis and regresses during the successive differentiation period. In some rare cases, it can persist in children and in adults, where it can be an occasional finding on radiological investigations or it can display insidious and non-specific symptoms due to progressive cerebrospinal fluid (CSF) accumulation and cystic enlargement of the cavity. The pathogenesis of VT persistency in adulthood and its cystic enlargement are still unclear and its management, whether conservative or surgical, is still an object of debate because of its rarity in the adult population.

Case presentation

A 52-year-old female patient with history of breast cancer, previously treated with chemo- and radiotherapy, presented with episodes of right leg clonus in the last

18 months, with hypoesthesia and weakness of the right thigh. Magnetic resonance imaging (MRI) of the spine showed an intramedullary cystic lesion at D10–D12 [length: 5.6 cm; volume: 6.6 cm³ (*Figure 1A,B*)], of which content was similar to CSF in both T1 and T2-weighted images without any post-contrast enhancement, and then reported as a terminal ventricle. No other pathological finding or spinal dysraphism at the lumbar and dorsal spine were evident. The patient was then addressed to conservative management and radiological follow-up.

After one year, a new MRI showed an increase of the cystic lesion [length: 6.4 cm; volume: 7.9 cm³ (*Figure 1C,D*)], and the patient reported a worsening in both the clonus and the weakness at her right leg. Thus, we decided to perform a cyst—subdural marsupialization through D10–D12 laminectomy and median myelotomy (*Figure 2A*). During the surgical procedure the cystic content came out under pressure through the myelotomy walls and an amount was collected for cytological evaluation that confirmed the CSF nature of the sample. The immediate post-operative period was uneventful and the patient was discharged on day four. The 3-month follow-up MRI showed a marked reduction of

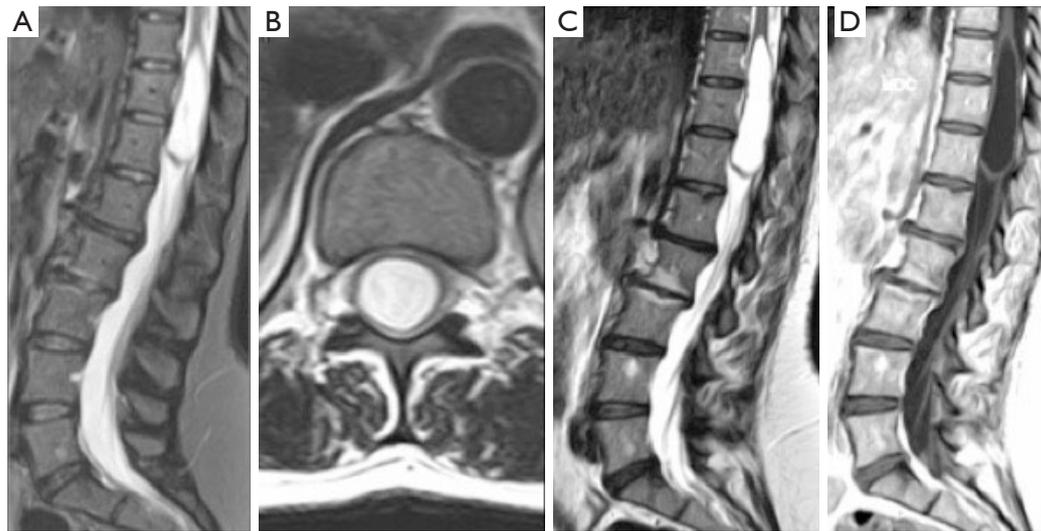


Figure 1 First (A,B) and 1-year-control (C,D) MRI of a 52-year-old female complaining of hypoesthesia and weakness of her right thigh. (A,B) Sagittal and axial T2-weighted MRI images showing an intramedullary ventriculus terminalis (length: 5.6 cm; volume: 6.6 cm³); (C,D) the 1-year follow up MRI showed an increase of the cyst dimensions (length: 6.4 cm; volume: 7.9 cm³), without post-contrast enhancement.

the dimension of the intramedullary cavity [length: 5.9 cm; volume: 2.9 cm³ (*Figure 2B*)] and the patient referred an improvement of the weakness and no further episodes of clonus.

Discussion

The ventriculus terminalis (VT), or ‘fifth ventricle’ (1), is an ependyma-lined space located in the conus medullaris, containing CSF and communicating with the central canal of the anterior portion of the spinal canal. It represents a normal phase of the development of the neural tube, appearing between the 43th and the 48th day of embryogenesis, and disappearing during the regressive differentiation period (1). Nevertheless, a persistent VT can be documented as an occasional finding in children less than 5 years old (2). In adults, it is a very rare finding with only 57 documented cases reported to date (*Table 1*).

The etiopathogenesis is still unclear; the persistence of the VT can be caused by vascular disturbances, inflammatory diseases, compressions, ischemic necrosis of the spinal cord (3) or due to an isolation of the VT from the central canal resulting from its dysembriogenetic failed regression (11), with a progressive cystic dilatation of the cavity (6).

Up to now, this is the second case reported of a terminal ventricle that showed a progressive enlargement in the radiological follow-up (22).

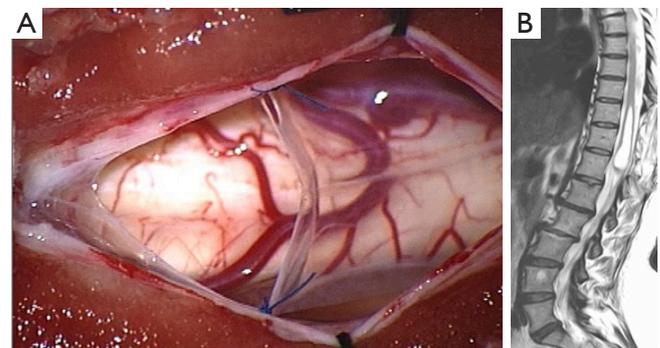


Figure 2 Microscopic view of the bulged spinal cord (A) and post-operative sagittal MRI (B). (A) Intraoperative image obtained after midline myelotomy and fenestration of the VT, allowing CSF leakage from the myelotomy walls and subsequent reduction of the swelling of the spinal cord; (B) 3-month post-operative T2-weighted MRI, showing reduction of the cavity (length: 5.9 cm, volume: 2.9 cm³). VT, ventriculus terminalis; CSF, cerebrospinal fluid.

Symptoms can be various, as some patients complain of non-specific symptoms (e.g.: low back pain, sciatica) and others exhibit focal neurological deficits, such as gait disturbances, paresis, muscular atrophy and sphincter dysfunction (13). The clinical presentation is also variable, ranging from an insidious onset (10,11) to a rapidly worsening form (6,8,10), even if only one case of an acute

Table 1 Literature review of reported cases of cystic dilatation of a ventriculus terminalis

Author and year	No. of patients	Treatment	Outcome
Nassar <i>et al.</i> , 1968 (3)	3	Surgical	Improved
Stewart <i>et al.</i> , 1970 (4)	3	Surgical	Improved
Korosue <i>et al.</i> , 1981 (5)	1	Surgical	Improved
Sigal <i>et al.</i> , 1991 (6)	4	Surgical [2], conservative [2]	Unknown
Agrillo <i>et al.</i> , 1997 (7)	1	Surgical	Improved
Matsubayashi <i>et al.</i> , 1998 (8)	2	Surgical	Improved
Celli <i>et al.</i> , 2002 (9)	1	Conservative	Stable
Dullerud <i>et al.</i> , 2003 (10)	2	Surgical	Improved [1], stable [1]
Liccardo <i>et al.</i> , 2005 (11)	1	Surgical	Stable
Brisman <i>et al.</i> , 2006 (12)	1	Surgical	Improved
de Moura Batista <i>et al.</i> , 2008 (13)	2	Surgical	Improved
Ciappetta <i>et al.</i> , 2008 (14)	2	Surgical	Improved
Takahashi <i>et al.</i> , 2009 (15)	4	Surgical [1], Percutaneous aspiration [3]	Improved
Borius <i>et al.</i> , 2010 (16)	1	Surgical	Improved
Dhillon <i>et al.</i> , 2010 (17)	1	Surgical	Improved
Ganau <i>et al.</i> , 2012 (18)	13	Surgical	Improved
Suh <i>et al.</i> , 2012 (19)	10	Surgical [4], conservative [6]	Improved [4], stable [6]
Bellocchi <i>et al.</i> , 2013 (20)	1	Surgical	Improved
Şimşek <i>et al.</i> , 2016 (21)	1	Unknown	Unknown
Woodley-Cook <i>et al.</i> , 2016 (22)	1	Surgical	Improved
Kawanishi <i>et al.</i> , 2016 (23)	1	Surgical	Improved
Present case	1	Surgical	Improved

cauda syndrome has been reported (12).

The diagnosis was based on the MRI that showed a cystic dilatation at level of the cauda, with smooth walls and without internal septae, communicating with the spinal central canal. The cystic fluid has a similar signal to CSF on all sequences (hypointense on T1-weighted, isointense on proton density weighted and hyperintense on T2-weighted images) and does not show post-contrast enhancement. These characteristics are useful to discern it from other more common lesions at this level such as syringohydromyelia, intramedullary cyst or glial tumors. MRI is also useful in detecting other cranial or spinal conditions, such as spinal dysraphisms, Chiari malformation or tethered cord syndrome that are rare comorbidities in VT patients.

Because of the rarity of VT and the variability of the

neurological condition in such patients, no standard treatment has been defined. Surgical management is represented by the fenestration of the cyst through laminectomy and a small midline myelotomy; after the drainage of the cavity the cyst is marsupialized to obtain a good CSF flow. This technique, similar to the one described by Nassar *et al.* (3), allows a complete recovery in 52% of cases and a partial recovery in 43% of cases (16). The placement of a shunt between the cyst walls and the subarachnoid space has been reported only in a few cases (4), albeit this procedure is still controversial because of a higher rate of complications and failure in the long-term follow-up. Recently, an alternative procedure of percutaneous aspiration of CSF during real-time MRI has been proposed with good results, but only in three patients (15).

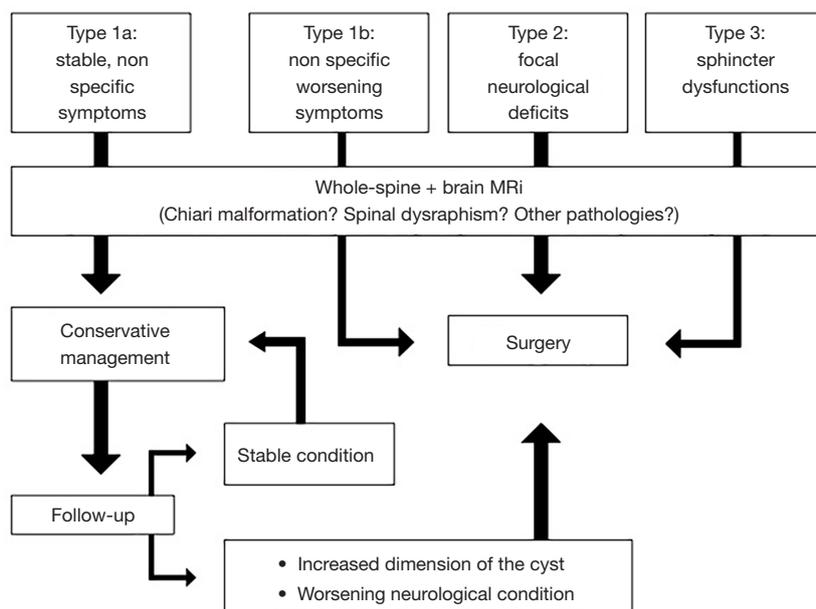


Figure 3 Proposed model of a management flow-chart for VT patients: patients with non-specific and stable symptoms should be managed conservatively. The surgical procedure should be reserved for patients with worsening symptoms or neurological deficits clearly related to VT (Types 1b, 2 and 3), or in Type 1a patients with dimensional increase of VT or worsening symptoms at the follow-up control. VT, ventriculus terminalis.

Because of these suboptimal outcomes, due to an empirical and subjective management, and considering that only patients with specific neurologic symptoms clearly related to VT showed a better outcome at the follow-up, de Moura B. *et al.* classified VT symptoms as three types and stated that surgical management should be reserved only in patients with neurological deficits or sphincter dysfunctions (Type 2–3), while conservative management, with a close neurological and radiological follow-up, should be preferred in patients with nonspecific complaints (Type 1) (13). Ganau *et al.* suggested a further subdivision of Type 1 symptoms into Type 1a, in which symptoms are non-specific and stable within a couple of months that can be managed conservatively, and Type 1b, with non-specific but worsening symptoms, requiring surgical management (18).

In our case the VT was originally an incidental finding because the first MRI was performed after the onset of clonus at her right leg and then treated surgically because of the dimensional increasing of the cyst and the worsening of the neurological condition. Before the surgical procedure the patient performed both brain and whole-spine MRI that did not show any other pathology (i.e., brain metastasis, Chiari malformation). The surgical drainage of the

cyst content followed by subdural marsupialization was successful and the patient reported an improvement of her neurological condition, with evidence of a dimensional reduction of the cyst at the post-operative MRI.

To date, there is a lack of a standardized guideline for VT patients even if multiple authors have proposed a management chart subdividing the patients according to their symptoms and argue that only patients with neurological deficits or worsening condition should be treated surgically. We agree with this principle but we suggest performing a complete pre-operative radiological evaluation of both the brain and the spine to exclude the presence of other pathologies (i.e., brain/spinal tumor; Chiari malformation). Furthermore, clinical and radiological follow-up criteria should be included in the management chart for patients with non-specific symptoms (Type 1a), as shown in *Figure 3*.

Conclusions

A VT should not be considered as a steady, non-pathologic anatomical anomaly. In our case a VT showed a progressive enlargement with worsening of the neurological condition of the patient. Therefore, in both symptomatic and

asymptomatic cases of fifth ventricle we suggest to perform a complete brain and spinal radiological investigation, in order to exclude other pathologies at these levels.

The surgical procedure of cyst fenestration through laminectomy and posterior median myelotomy is more advisable than a percutaneous aspiration of the cyst that has been described only for three patients and represents, in our opinion, a hazardous procedure due to a higher risk of damaging the rich perimedullary vascular network.

The rarity and unclear pathogenesis, together with the absence of a standard management and its unpredictable behaviour in adults had led to suboptimal results in VT patients, especially for those with non-specific neurological symptoms. We therefore stress the importance for further clinical studies and long term follow-up in operated patients to better define a univocal guideline for the management of this uncommon condition.

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None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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