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Letter to the editor

Surgery for cyst dilatation of a ventriculus terminalis in adults: Keep it simple!

Dear editor,

Ventriculus terminalis (VT) is defined as an ependymal cell lined cavity within the conus medullaris and containing cerebrospinal fluid (CSF) [1–3]. This is considered as an anatomical variance up to the age of 5 and has been classically described on ultrasonography during the neonatal period [4]. The etiology of such cystic dilatation is debatable. While some authors consider VT cysts as a developmental defect (appearing between the 4th and 6th intrauterine life), others report trauma, vascular or compression as the main cause [3,5].

In normal adults, the VT can only be demonstrated on histology, as it is not routinely seen on imaging [6]. The diagnosis is so usually incidental. They are frequently asymptomatic, especially in children. However, symptoms may appear due to the cystic dilatation itself with further compression of the nerve tissue in conus medullaris.

The management of rare cyst dilatation of VT, with less than 30 cases reported in the literature, is not well established [1]. Conservative management, marsupialization, or placements of a "T" drain have been described. We describe our experience in 3 cases in this rare pathology [7]. We share our surgical indications and describe our operative approach.

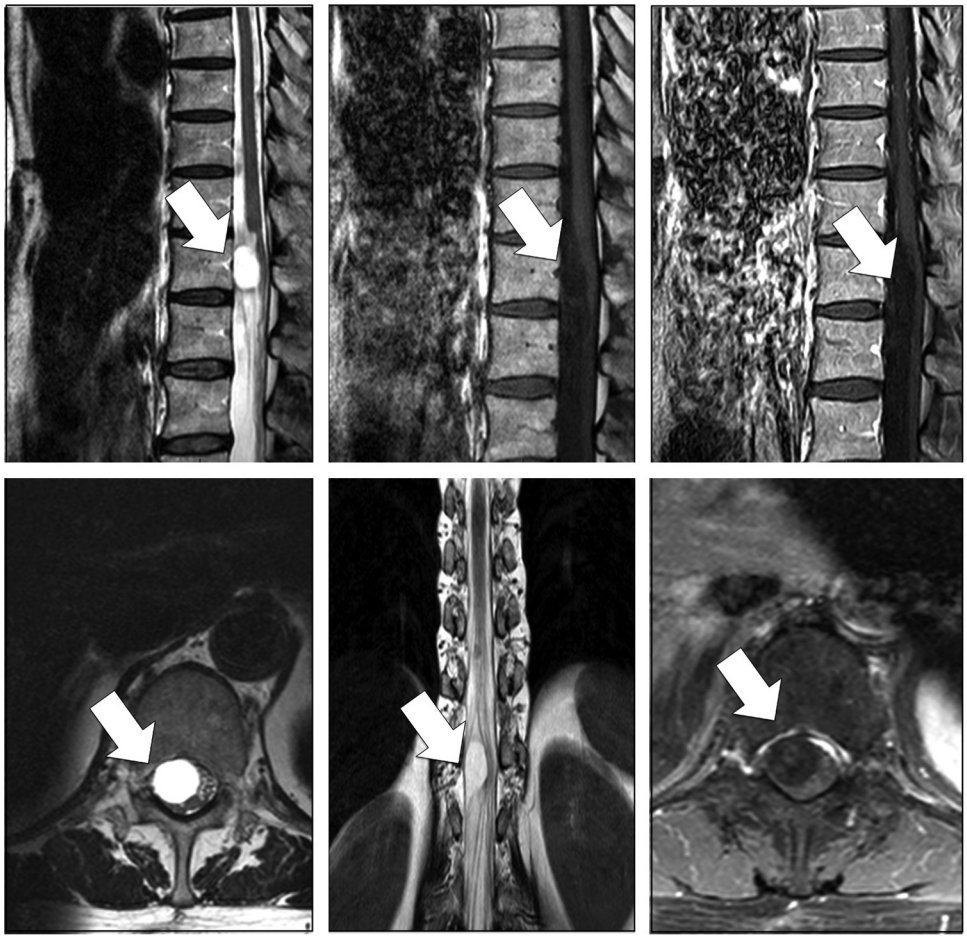

Fig. 1. Illustrative case 1 with preoperative MRI in sagittal, axial and coronal plane, while exemplifying different sequences.

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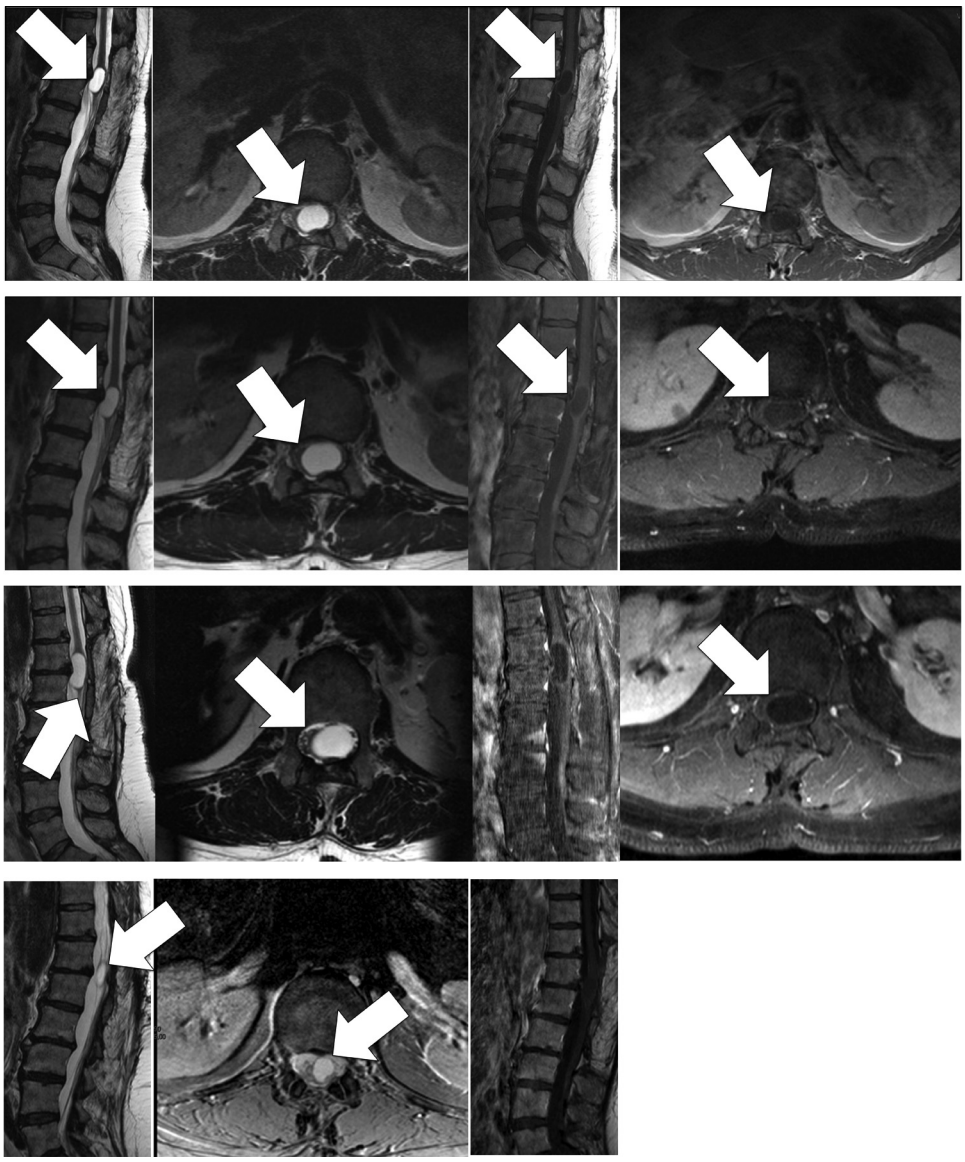

Fig. 2. Illustrative case 2 with preoperative MRI in sagittal and axial plane, while demonstrating different MR sequences at 2 years (2.1.), 1 year (2.2.) and 6 months (2.3.) preoperatively and 18 months after second surgery (2.4.), respectively.

1. Case series presentation

1.1. Case #1

A 70-years-old female patient presented with progressive motor deficit during the past 3 years, mainly at the levels L5 left, L5 right and S1 right. MRI revealed cystic dilatation of the VT at the level of T12 (Fig. 1). The cavity was hypointense in T1 and hyperintense in T2, fulfilled with fluid with the same signal as the CSF. She underwent surgical exploration with laminectomy and marsupialization, which was uneventful. Postoperative clinical examination showed complete motor recovery.

1.2. Case #2

A 55-years-old female patient had been previously operated for a cystic dilatation of the VT at the levels T12 thirteen years back. During the following 10 years after surgery, she presented with clinical and radiological uneventful course. Afterwards, she presented with left lower leg weakness and progressive sciatica. MRI revealed progressive increase of the VT cyst's volume (Fig. 2). Laminectomy and further cyst marsupialization were performed. Postoperative clinical examination revealed full recovery of the motor symptoms. Postoperative MRI showed major decrease in size, which persisted up to 5 years after the second surgery, date of the last follow-up of the patient.

1.3. Case #3

A 57-years-old female patient presented with left leg pain during the past 3 years and, more recently, sacral pain. During the past months, she also developed a medullary claudication. Clinical examination revealed a bilateral L5 motor root deficit. Preoperative MRI showed a cystic dilatation of the VT at the levels T11-T12-L1. She underwent surgical exploration with T12-L1 laminectomy, myelotomy, and dissection of local arachnoiditis and placement of a shunt fixed to pia mater. Postoperative clinical course was marked by complete motor recovery.

2. Discussion

Ventriculus terminalis cyst dilatations are rare and challenging cases. Asymptomatic patients with no radiological progression can benefit from regular clinical and radiological follow-up. Symptomatic cases with posterior cysts can be offered laminectomy, myelotomy and marsupialization. In patients presenting with additional focal arachnoiditis, which might be subject of further cyst recurrence, a "T" shunt can be placed and further attached to pia mater.

Physiological role of VT is currently undiscovered. The most common theory places VT at the cross point between ending of Reissner fibers (which extend from subcomisural organs of the epithalamus to VT), while storing neurosecretory substances. Their potential pathophysiological functions are mainly of mechanoreception, to indicate variations in CSF pressure. Furthermore and by this, they can regulate the quality of CSF.

An isolated rare VT is not typically associated with dysraphic pathologies and is usually seen during fetal development, while being present in vast majority of cases after birth. Before the age of 5 is considered normal anatomical variance, with propensity to further regress [4,8]. In persistent asymptomatic cases, other abnormalities of craniospinal axis, such as spinal cord tethering, tumors, etc. should be excluded. In other associated particular conditions, such as syringomyelia, dilatation is usually present in upper parts of spinal cord and is frequently associated with Chiari type I, myelomeningocele, etc. [9]. Central canal is normally broadest at conus medullaris level and if prominent or enlarged, any cystic neoplasm or syringohydromyelia should be formally excluded.

The exact origin of clinical and radiological decompenssation in adults of cyst enlargement of VT remains unclear. Some authors have hypothesized that inflammatory processes (such in our case #3), medullary ischemia, medullary compression or trauma could interfere and create a lack of communication between the VT and the ependymal canal [2,3,5,6]. Pathophysiologically, this engenders an altered CSF flow within central canal or leads to disturbance of the Reissner Fiber (a subependymal secretory product), which has a role in CSF regulation [10].

MRI plays the role of standard paraclinical assessment. The former classically shows anatomical location at the level of the conus medullaris, T12-L2, with a cystic cavity presenting with same signal as the CSF, and no signs of solid portion. Furthermore, there is no evidence of tumor signs and no contrast enhancement. Further investigations might include electromyography examination, lemniscal conduction or urodynamic assessment, shall urinary symptoms be present.

Recurrence is unusual and can be further treated by new surgical exploration. In this context, placement of a "T" drain might be also discussed.

Compliance with ethical standards

Yes.

Ethics approval

Ethical Committee Approval has been required, according to the French rules, at the *Assistance Publique*, *Hopitaux de Paris*, Paris, France, for this retrospective review of cases.

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Disclosure of interest

The authors declare that they have no competing interest.

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