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ORIGINAL RESEARCH

Treatment Strategies for Unknown Anatomic Pathology Ventriculus Terminalis Case Series: Single Center Experience*

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ABSTRACT

The ventriculus terminalis is a cavity in the conus medullaris, bounded by ependymal cells, associated with the central canal. It is an anatomical structure that is very rare in adults, with a limited number of surgical cases that have been reported in the literature. In children, it is regarded as a normal congenital variation, known to regress before five years of age, and very few symptomatic cases have been reported in both pediatric and adult populations. It is often asymptomatic in adults and is detected incidentally. Although potentially nonsignificant individually, symptoms can range from nonspecific low back pain to sphincter dysfunction and focal neurologic deficits. Our purpose is to discuss our management strategy in comparison to the existing literature. A retrospective review was conducted of all adult patients (aged 17 years and older) diagnosed with ventriculus terminalis who were referred to the hospital between 2010 and 2020. Clinical classification was made according to the classification defined by Batista. In addition, Ganau's classification was also used. Five patients were included in the study. The majority of these patients (n=4, 80%) were symptomatic at the time of diagnosis, with nonspecific back pain being the most common symptom (n=3, (60%). None of the patients required neurosurgical intervention during the follow-up period of 21.6±8.9 months, as there was no clinical deterioration observed. Ventriculus terminalis is a rare pathology that may develop de novo in adults, often remaining undiagnosed until the cyst enlarges, and can manifest with a wide spectrum of symptoms. When identified, it requires careful management, involving surgery when necessary and a conservative approach when appropriate.

Keywords: Ventriculus terminalis. Cauda equine. Fifth ventricle. Spinal cord.

Bilinmeyen Anatomik Patoloji Terminal Ventrikül Tedavi Stratejileri: Tek Merkez Deneyimi

ÖZET

Terminal ventrikül, konus medullariste bulunan, ependimal hücrelerle sınırlanan, santral kanalla ilişkili bir boşluktur. Literatürde sınırlı sayıda cerrahi yapılan olgu bildirilen, erişkinlerde oldukça nadir görülen bir anatomik yapıdır. Çocuklarda, beş yaşından önce gerilediği bilinen normal bir konjenital varyasyon olarak kabul edilmektedir ve hem pediatrik hem de yetişkin popülasyonda çok az sayıda semptomatik vaka rapor edilmiştir. Yetişkinlerde sıklıkla asemptomatiktir ve tesadüfen tespit edilir. Semptomlar nonspesifik bel ağrısından sfinkter fonksiyon bozukluğuna ve fokal nörolojik defisitlere kadar değişebilir. Amacımız nadir görülen bu patolojideki yönetim stratejimizi mevcut literatürle karşılaştırmalı olarak tartışmaktır. 2010-2020 yılları arasında hastaneye başvuran terminal ventrikül tanısı alan tüm erişkin hastalar (17 yaş ve üzeri) retrospektif olarak incelendi. Klinik sınıflandırma Batista'nın tanımladığı sınıflamaya göre yapıldı. Ayrıca Ganau'nun sınıflandırması da kullanıldı. Çalışmaya 5 hasta dahil edildi. Bu hastaların çoğunluğu (n=4, %80) tanı anında semptomatik olup, en sık görülen semptom nonspesifik sırt ağrısıydı (n=3, (%60). Hiçbir hastanın kliniğinde kötüleşme olmadı, takip süresi 21,6±8,9 aydı. Terminal ventrikül erişkinlerde de novo olarak gelişebilen, sıklıkla kist büyüyene kadar tanı konulamayan ve geniş bir semptom yelpazesiyle ortaya çıkabilen nadir bir patolojidir. Saptandığında, gerektiğinde cerrahiyi ve uygun olduğunda konservatif bir yaklaşımı içeren dikkatli bir tedavi gerektirir.

Anahtar Kelimeler: Terminal ventrikül. Kauda equina. Beşinci ventrikül. Spinal kord.

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According to the accepted views and publications in the literature, the 5th ventricle is defined as the expansion of the central canal at the conus level[1]. It is seen that in communicating hydrocephalus, the central canal can expand like a ventricle and therefore it is called the 5th ventricle in this terminology[1,2]. According to Zeinali et al., this entity was first described by Stilling in 1859 and followed by Krause in 1875[3]. It was later recognized as a true fifth ventricle by Kernohan in 1924[4]. The ventriculus terminalis (VT) is a cavity located in the conus medullaris, bounded by ependymal cells and associated with the central canal[3]. According to the existing literature, VT is an ependymal-lined cavity formed through canalization and retrogressive differentiation during embryological development[3-5].

Key features of this cyst include a smooth-walled, predominantly oval contour found exclusively in the conus medullaris, displaying a hyperintense signal on T2-weighted magnetic resonance imaging (MRI) and a hypointense signal similar to that of cerebrospinal fluid on T1-weighted MRI. Furthermore, no contrast enhancement and septation are observed after contrast agent injection[5,6-10]. The differential diagnosis for VT includes tumoral and space-occupying lesions like hydrosyringomyelia, ependymoma, astrocytoma, and hemangioblastoma[5,9]. Rarely, VT may also coexist with congenital malformations such as split cord, bifid spine, and dermoid sinus tract[11,12].

Asymptomatic localized dilatation of VT is a normal developmental phenomenon that occurs in approximately 2.5% of children under five years of age[12,13]. Radiological progression is rare, with only five cases reported so far[10,14,15,16]. Surgical treatments for VT include cyst fenestration[5,8-10,12,15-23], puncture, and catheter-assisted cyst-subarachnoid shunt[6,8,16,21] procedures. Here, we aim to discuss our management strategy compared to the existing literature.

Material and Method

A retrospective review was conducted on all adult patients (aged 17 years and above) diagnosed with ventriculus terminalis and referred to the hospital between 2010 and 2022. Medical records and imaging data from digital hospital charts were retrospectively reviewed. This comprehensive analysis encompassed patient demographics, neurological examination findings, and radiological imaging assessments. As mentioned before, radiologically, oval lesions with cystic expansion that did not show any contrast enhancement at the conus level were considered to be VT and were included in the study. The median cyst volume, as measured using the formula (length × width × height)/2[16]. Clinical classification was made according to the classification defined by Batista[18]. In addition, Ganau's classification was also used[8]. The patients were followed-up for an average period of 21.6±8.9 months.

Results

Five patients with MRI-verified VT were included in the study, comprising four females (%80) and one male (%20). The mean age of the patients was 35.6±10.3 years (range 17-47 years). The diagnosis of VT was established through radiological examinations. In three patients, the diagnosis was prompted by radiological studies conducted in response to nonspecific low back and back pain, while in one patient, it was discovered during metastasis scans related to breast cancer. The remaining patient was under observation for transverse myelitis, and VT was incidentally detected during radiological investigations following acute paraplegia (Table I). All lesions were at the conus level, and the cyst dimensions were calculated as 5.7±10.2 (range 0.1-28) ml (Figures 1a-h and 2a-f). No patient in our series had a spinal space-occupying lesion or congenital spinal dysraphism. No Chiari malformation was detected in the cranial MRI examination performed for other reasons in 3 patients. Our other 2 patients did not have any Chiari symptoms or signs.

Table I. Characteristics of the patients with ventriculus terminalis

Patient	Age	Sex	Complaint	NE	Cyst Volume (ml)	Progression
1	43	F	None	WNL	4x4x2	None
2	47	F	Back pain	WNL	24x18x13	None
3	37	F	Back pain	WNL	20x2x2	None
4	17	F	Leg weakness	Paraplegic	24x8x6	None
5	34	M	Back pain	WNL	14x9x10	None

F: Female, M: Male, NE: Neurological examination, WNL: Within normal limits

All patients were categorized as type 1 VT cases and received conservative treatment. In the patient where paraplegia regressed after pulse steroid and rehabilitation, surgical intervention was not deemed necessary. Subsequent follow-up assessments showed no signs of progression or clinical worsening during a follow-up period of 21.6±8.9 months.

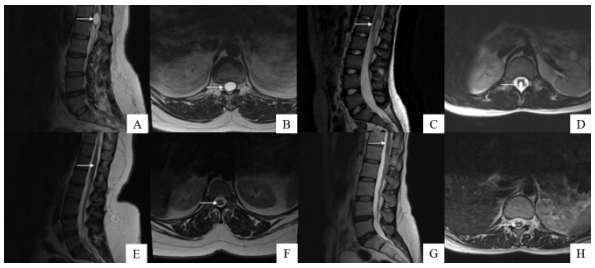

Figure 1.
Radiographic Features: A, B; C, D; E, F and G, H show the sagittal and axial T2 MRI images of the patients. White arrows indicate the terminal ventricle.

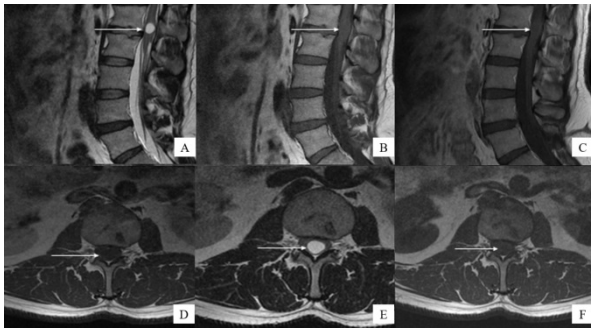

Figure 2.
VT Characteristic Radiological Features: The white arrows show the cyst located in the conus. In the images of patients A, B, and C, sagittal T2, T1 and contrast-enhanced T1, D, E, and F axial T2, T1 and contrast-enhanced T1 Lumbar MRI images. T2 images show a hyperintense lesion and T1 and contrast-enhanced T1 images show a lesion that is isointense with cerebrospinal fluid, with no contrast enhancement.

Discussion and Conclusion

VT is an exceedingly rare condition in adults, often presenting as a *de novo* occurrence, with fewer than 100 surgical cases documented in the medical literature. In contrast, it is considered a normal congenital variation in children, generally regressing before age five. Symptomatic cases are exceptionally scarce in both pediatric and adult populations. In adults, VT is often asymptomatic and typically discovered incidentally. A study by Suh et al. concluded that there is no pulsatile movement within the cyst by examining the dynamics of intra-cyst cerebrospinal fluid[7]. Neuroradiological techniques can visualize VT in adults only when it is dilated[12]. Tancioni et al. were the first to report the *de novo* formation of VT; however, the mechanism behind this formation remains unexplained[9]. Considering the onset of symptoms in all our cases in our series, it can be thought that this pathology emerged *de novo*. However, we do not have any childhood imaging to prove otherwise.

It often eludes diagnosis until the cyst reaches a certain size and can manifest with a wide range of symptoms, spanning from nonspecific low back and back pain to sudden onset of cauda equina syndrome. Despite its elusive nature, most VT cases are observed in female patients[3]. In our series, there was a patient group, mostly female.

Cystic enlargement of this structure can manifest with a wide array of clinical presentations with symptoms ranging from nonspecific low back pain to sphincter

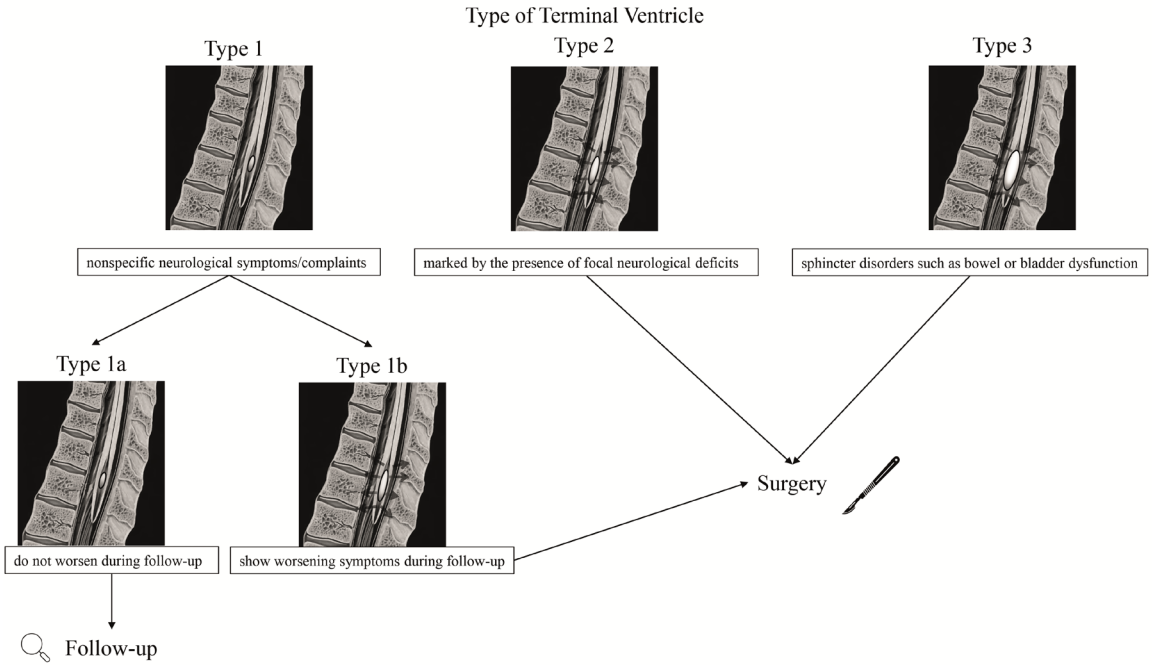

Figure 3.
Follow-up and treatment management are summarized with a simple illustration.

dysfunction and focal neurologic deficit[3,5,14,17]. In our series, the diagnosis was made in 4 patients during examination of symptoms and findings (80%) and in one patient incidentally (20%). The pathophysiology is still not elucidated. The VT, the distal end of the Reissner fiber arising from the epithalamus, has been hypothesized to function as a mechanoreceptor sensitive to changes in cerebrospinal fluid pressure[24]. It is conceivable that cyst size is associated with neurological symptoms and worsening. In our series, cyst sizes were significantly lower in two patients (0.1 and 0.4 ml) and symptoms resolved quickly with conservative treatment. Although the cyst sizes of the other patients were larger than the other two patients, the fact that there was no worsening and no radiological progression during follow-up may be an indication that this disease is stable.

Based on Batista's clinical classification, VT is categorized into three types: VT type I, characterized by nonspecific neurological symptoms or complaints; VT type II, marked by the presence of focal neurological deficits; and VT Type III, where sphincter disorders such as bowel or bladder dysfunction are observed[18]. In their series, Ganau et al.[8], in addition to Batista's classification, suggested examining type 1 in two subgroups: those that do not worsen during follow-up (Type 1a) and those that show worsening symptoms during follow-up (Type 1b). Accordingly, surgery is considered the gold standard for type 2 and type 3 VT cases, while the optimal approach for Type 1 cases remains uncertain. In fact, Type 1 cases often lean towards conservative treatment[5-8,17]. Although the Type II and Type III classifications have a clear surgical indication[8,18,19], introducing the Type Ia and Ib subgroups may help establish a new guideline for an appropriate surgical indication[8]. In our series, all cases remained stable in the following months and did not require any surgery. In this case, it would be correct to include the cases in Type 1a in the classification proposed by Ganau (Figure 3). There are very few series in the literature with conservative treatment, and our study is one of the series that reached the highest number of patients[7,8,24].

In adults, only 68 surgically treated cases have been reported in the literature[10]. In a case series published by Fletcher, one of the 12 surgically treated patients experienced recurrence, prompting that shunting could be beneficial in reducing recurrence for cysts[16]. However, it is worth noting that patients who underwent cyst-subarachnoid shunt procedures faced a notable risk of shunt complications and secondary recurrence due to shunt dysfunction. According to a review published in the literature, which is the most current and comprehensive, a total of less than 100 surgical series were comprehensively evaluated[24]. According to this review, it has been reported that the 5th ventricle can be diagnosed quickly and effectively with a multidisciplinary approach, conservative treatment may be beneficial in selected cases, and in case of neurological deterioration, the results of early surgery are good, and symptoms worsen only in a small group of patients[24]. Many issues need to be investigated about the terminal ventricle because the cases are in the form of small case series, lack of long-term follow-up, and heterogeneity of the study groups.

In our clinical experience, we encountered a case in which a neurological deficit coincided with another pathology. In this instance, a conservative approach was pursued. The remaining patients showed no further deterioration during their clinical and radiological follow-ups. When identified, it should be managed appropriately, which may involve surgery when necessary or a conservative approach when suitable. For type 1a cases, conservative follow-up has proven to be both safe and effective. In other instances, treatment decisions may be tailored to the individual patient's needs. The issue that needs to be considered in long-term follow-up is neurological deterioration or progression of symptoms. This problem can be overcome if patients are well informed on this issue and have easy access to healthcare services. In necessary cases, information or follow-up cards can be used to ensure that the patient comes for an annual follow-up even if he has no symptoms. The point that should not be forgotten is that the classifications are clinically-based, and patient-specific evaluation is important in making a surgery or follow-up decision.

The limitations of this study were that a small number of cases were examined and there was no long-term follow-up. The fact that only 5 patients were observed with this pathology during a 10-year period may indicate how rare this disease appears to be. Additionally, since it is an unknown pathology, it should be remembered that there may be asymptomatic cases that remain undiagnosed, and the true incidence can be determined through more comprehensive studies.

These cases should be recognized early with a multidisciplinary, as they can lead to rapid neurological deterioration, early surgery can give good results, and conservative treatment can be successful in selected cases. Clinical, anatomical and neurophysiological studies with larger case series and long-term results are needed for this rare and still unknown pathology.

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