Case Report

A RARE CASE OF KRAUSE TERMINAL VENTRICLE WITH LOW-DISPLACEMENT MEDULLARY CONE AT L4

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ABSTRACT

The terminal ventricle (TV) of Krause is a rare cystic dilation of the central canal of the medullary cone. Due to the limited understanding of its pathophysiology, the optimal management of this condition remains a subject of ongoing discussion. In this article, we report the case of a 43-year-old male who presented with stabbing pain at the thoracolumbar junction for approximately two months. Magnetic resonance imaging of the spine revealed a small fusiform dilation of the ependymal central canal, with a Krause terminal ventricle measuring a maximum transverse diameter of 0.5 cm, located at the level of the medullary cone, which exhibits a low displacement at the L4 level.

KEYWORDS: *fifth ventricle, terminal ventricle, Krause's ventricle, ventriculus terminalis, Fedor Krause*

INTRODUCTION

The terminal ventricle (TV), also known as Krause's terminal ventricle, is a transient ependymal cystic cavity that develops in the medullary cone, which is the lower portion of the spinal cord (1, 2). Initially considered a vestigial remnant of the embryonic neural tube, the TV may persist in a variable percentage of adult individuals (1-3). First described by Stilling in 1859 and later renamed the "fifth ventricle" by Krause in 1875, the TV represents a dilation of the central canal in the medullary cone. This process originates during embryogenesis in the caudal section of the central canal of the neural tube and typically regresses spontaneously in early childhood (1-3). However, in some cases, the TV may persist or develop abnormally, sometimes leading to neurological symptoms. Although the condition is often asymptomatic, its pathogenesis and clinical significance remain subjects of ongoing discussion. Rarely, cystic expansion of the TV can compress surrounding neural structures, causing symptoms such as lower back pain, bladder and bowel dysfunction, and neurological deficits in the lower limbs (1-4).

The diagnosis of TV is primarily made through Magnetic Resonance Imaging (MRI), which serves as the reference diagnostic method for this condition, allowing for an accurate evaluation of its morphological characteristics (5-7). It is crucial to adopt a differential diagnostic approach to exclude other cystic pathologies that may affect the spinal cord, such as syringomyelia, arachnoid cysts, and malignant neoplasms (8). The treatment of TV depends on the severity of symptoms and the potential for progressive neurological deterioration. In some cases, symptoms may remain limited to nonspecific manifestations, while in others, more severe neurological complications may develop. The variability in symptoms complicates the establishment of a standardized treatment, with approaches ranging from periodic surveillance to surgical intervention, based on the severity of the condition and the individual characteristics of the patient (1-10).

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Due to the rarity of TV and the variability of the neurological condition in these patients, no standard therapeutic strategy exists. The primary surgical treatment involves cyst fenestration through laminectomy and small median myelotomy, followed by cavity drainage and cyst marsupialization to promote adequate cerebrospinal fluid (CSF) flow. This method, akin to the one outlined by Nassar et al. (5), resulted in complete recovery in 52% of cases and partial recovery in 43% (4, 11). In certain instances, a shunt has been placed between the cyst and the subarachnoid space, though its use remains controversial due to a higher incidence of complications and long-term failure. More recently, a new approach involving percutaneous aspiration of cerebrospinal fluid under real-time MRI guidance has been suggested, yielding promising outcomes, although it has only been tested in a small cohort of three patients (4, 12).

In an effort to optimize the management of TV, Moura et al. proposed a classification of symptoms associated with TV, dividing them into three main categories. According to this classification, only patients presenting with neurological deficits or sphincter dysfunctions (types 2 and 3) should undergo surgical treatment, while those with nonspecific symptoms (type 1) are recommended to receive a conservative approach, with continuous neurological and radiological monitoring (4, 9). Subsequently, in 2012, Ganau et al. updated this classification (13). The revised version distinguishes four types of terminal ventricle cystic lesion (CLVT): type Ia, which includes stable and nonspecific symptoms, with no clear correlation to the TV; type Ib, characterized by nonspecific symptoms that tend to worsen over time; type II, in which focal neurological deficits manifest; and type III, involving sphincter disturbances. According to this classification, patients with type Ia should be treated conservatively, while patients with types II and III require surgical intervention to alleviate neurological symptoms and prevent additional complications (6, 13-15).

The aim of this study is to present a case report of a 43-year-old man who came to the attention of the authors, presenting with stabbing pain at the thoracolumbar junction for approximately two months, with the goal of contributing to the scientific community's awareness of the etiology and characteristics of this clinical condition.

CASE REPORT

We report the case of C.A., a 43-year-old male presenting with stabbing pain at the thoracolumbar junction for approximately two months. Initially, the referring physician suspected a possible renal origin for the symptoms and requested an abdominal ultrasound, which yielded negative results. Given this outcome and the persistence of the painful symptoms, the physician decided to have the patient undergo a spinal MRI. This examination of the spine revealed a small fusiform dilation of the ependymal central canal, consistent with a Krause terminal ventricle. The ventricle had a maximum transverse diameter of 0.5 cm and was located at the level of the medullary cone, at the L3-L4 junction. The terminal ventricle exhibited signal hyperintensity on T2-weighted sequences, consistent with cerebrospinal fluid content, with well-defined margins and regular contours. Additionally, a thin fusiform dilation of the ependymal canal extending cranially from L4 to L3 was detected (Fig. 1A-D).

This case highlights the potential clinical implications of a persistent TV, which, although often asymptomatic, can sometimes lead to significant pain symptoms.

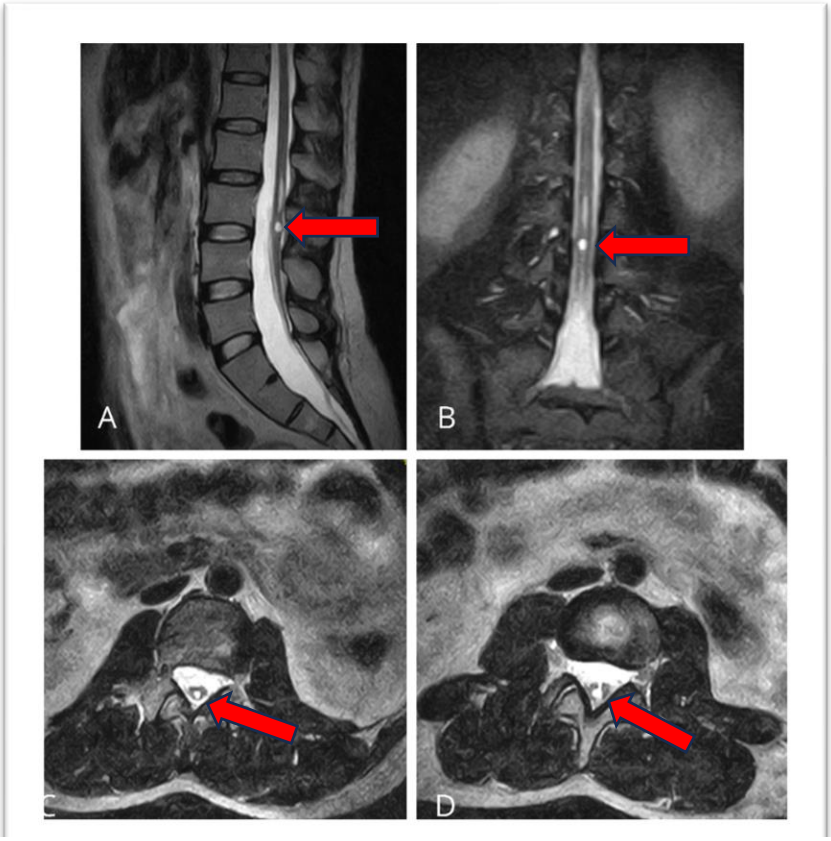

Fig. 1. *MR lumbar spine. **A**): Sagittal T2-weighted MR; **B**): T2-weighted MR; **C-D**): coronal and axial images. The MRI investigation reveals the presence of a fusiform dilation of the ependymal canal from L3 to L4, terminating with a small cystic dilation with a maximum transverse diameter of 0.5 cm, consistent with a Krause terminal ventricle (**arrows**).*

DISCUSSION

The Krause TV is a rare and transient cystic structure of the central canal of the medullary cone. Its prevalence and clinical significance are still subjects of debate, with the condition remaining asymptomatic in most cases. However, some patients may develop neurological symptoms, primarily due to compression of surrounding structures, which can lead to functional disturbances and lower back pain. Our clinical case of a 43-year-old man with stabbing lumbar pain represents an example of the potential clinical manifestations of a persistent TV.

The diagnosis of TV is typically made through MRI, which allows for the identification of dilation of the ependymal central canal and differentiation from other cystic pathologies that may involve the spinal cord, such as syringomyelia or arachnoid cysts (1, 8). In our case, the MRI revealed a small fusiform dilation of the central canal, consistent with a Krause terminal ventricle, located at the L3-L4 junction, confirming the presence of a condition that could explain the patient's symptoms. The progression of the TV can lead to painful symptoms, as in the case of our patient, who reported persistent pain at the thoracolumbar junction.

The management of TV depends on the severity of the symptoms and the potential for neurological progression. Most patients with asymptomatic TV do not require treatment, but patients with neurological symptoms or sphincter dysfunctions require surgical intervention (9). Surgical options include cyst fenestration through laminectomy, myelotomy, and drainage of the cystic cavity, followed by marsupialization to ensure proper cerebrospinal fluid flow (5, 11). The surgical technique is generally effective in reducing neurological symptoms, with a significant percentage of patients experiencing complete or partial improvement. However, despite positive outcomes, there are still risks of complications and recurrences, and surgical management remains controversial in some cases due to potential long-term side effects, as highlighted by our case.

The classification proposed by de Moura Batista et al. (9) and subsequently revised by Ganau et al. (13) provides a useful framework for guiding the management of TV based on the patient's symptoms. According to this classification, only patients with neurological deficits (type II) or sphincter dysfunctions (type III) should undergo surgical treatment. Those with nonspecific symptoms (type I) can be monitored conservatively, as in the case of our patient. However, the variability of symptoms and the risk of progression in patients with nonspecific symptoms necessitate regular neurological and radiological follow-up. The classification also distinguishes between stable and progressive symptoms in type I, suggesting that surgery may be required if symptoms worsen, as indicated by Ganau et al. (13-15).

Our case also highlights the need for a differentiated approach to managing patients with TV. Although most patients remain asymptomatic, the risk of painful symptoms or neurological dysfunctions must be carefully monitored, as TV can evolve and impact the patient's quality of life. Additionally, it is important to consider that the cystic expansion of TV may be accompanied by nonspecific symptoms, such as the thoracolumbar pain observed in our case, which complicates early diagnosis and appropriate management.

In conclusion, although Krause's terminal ventricle is a rare and often asymptomatic condition, its presence in certain patients can lead to significant clinical issues. Early diagnosis and appropriate monitoring, combined with the adoption of personalized therapeutic strategies, are crucial for preventing the progression of the condition. Conservative management is preferable for patients without severe symptoms, while surgical intervention is recommended for those who develop neurological deficits or sphincter dysfunctions. Further clinical studies and long-term follow-up are needed to refine therapeutic guidelines and improve the management of this condition.

CONCLUSIONS

Krause's TV is a rare condition that, although frequently asymptomatic, can present with significant clinical symptoms, as in the case of our patient with persistent lumbar pain. While most cases remain asymptomatic or exhibit only mild symptoms, early diagnosis through imaging techniques, such as MRI, is crucial for proper management.

The therapeutic strategy varies depending on the severity of symptoms. In asymptomatic patients or those with mild symptoms, a conservative approach with neurological and radiological monitoring is recommended. However, in cases where significant neurological symptoms occur, such as motor deficits or sphincter dysfunction, surgical intervention is indicated. Surgical techniques, such as cyst fenestration, have demonstrated a high success rate; however, they are not without risks, and patients require regular postoperative monitoring.

The symptom classification proposed by de Moura Batista et al. and later revised by Ganau et al. (13) provides a useful guide for determining when to intervene surgically and when to adopt a conservative approach. However, the

variability of symptoms and the potential for disease progression necessitate individualized assessment and ongoing follow-up.

In summary, while Krause's TV is a rare condition, it can have significant clinical implications. Timely diagnosis and a targeted therapeutic approach are crucial for preventing neurological complications and enhancing the patient's quality of life. Conservative management remains the primary option for asymptomatic cases, while surgical intervention is recommended for those with severe neurological symptoms. Further long-term studies are necessary to deepen the understanding of pathogenesis and optimize therapeutic guidelines.

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