Symptomatic Pediatric Ventriculus Terminalis, a Rare Ependymal-Lined Cavity of the Conus Medullaris: literature review

Ventriculus Terminalis Síntomático em Paciente Pediátrico uma Rara Cavidade do Cone Medular Revestida por Células Ependimárias: revisão de literatura

Lorimer Sandoval Carneiro¹
Lissa Carrilho Goulart¹
Tiago Vinicius Silva Fernandes¹
Caio Rodrigues Gomes Dias²
Julia Jordana Freitas Lima²

ABSTRACT

Introduction: The fifth ventricle, also known as ventriculus terminalis, is a term used to describe a cystic ependymal-lined cavity of the conus medullaris. Its persistence is a rare entity into adulthood and, in childhood, the ventriculus terminalis usually does not persist for more than 5 years of age. Methods: The literature review was performed in 2021 in the Medline (PubMed) and Lilacs (via BVS) databases, from the period after 1991. Articles that presented new cases of ventriculus terminalis in a population older than 5 years old, especially in the pediatric age group, diagnosed by ultrasound or MR of the dorsal and lumbar spine. Case Presentation: Male patient, 9-year-old, presenting episodes of paresis and bladder dysfunction due to a fifth ventricle at the level of T11-T12. Surgery was performed with good neurological outcome. Postoperatively, the patient presented CSF leak, which was corrected, surgically. Conclusion: An updated literature review is presented aiming to draw attention to a rare case of symptomatic ventriculus terminalis and to emphasize the importance of including the fifth ventricle in differential diagnosis of the spinal cord lesions to ensure proper treatment.

Keywords: Cyst fenestration; Conus medullaris lesions; Fifth ventricle; Ventriculus terminalis

RESUMO

Introdução: O quinto ventrículo, também conhecido como ventriculus terminalis, é um termo usado para descrever uma cavidade cística revestida pelo epêndima do cone medular. Sua persistência é uma entidade rara na idade adulta e, na infância, o ventriculus terminalis geralmente não persiste por mais de 5 anos de idade. Métodos: A revisão da literatura foi realizada em 2021 nas bases de dados Medline (PubMed) e Lilacs (via BVS), do período posterior a 1991. Seleccionamos os artigos que apresentavam casos de ventriculus terminalis em uma população maior de 5 anos de idade, principalmente na faixa etária pediátrica, diagnosticados por ultrassonografia ou ressonância magnética da coluna dorsal e lombar. Relato do Caso: Paciente de 9 anos com episódios de paresia e disfunção urinária devido ao ventriculus terminalis localizado ao nível de T11-T12, submetido a laminectomia e fenestração cirúrgica da dilatação, com boa evolução neurológica. No pós-operatório, o paciente apresentou fístula líquórica, a qual foi corrigida cirurgicamente. Conclusão: Uma revisão atualizada da literatura no intuito de chamar a atenção para um caso raro de ventriculus terminal sintomático e enfatizar a importância da inclusão do quinto ventrículo no diagnóstico diferencial das lesões medulares, para garantir o mais adequado tratamento.

Palavras-Chave: Fenestração cística; Lesões do cone medular; Quinto ventrículo; Ventriculus terminalis

¹ MD, Division of Neurosurgery, Department of Surgery, Medical School, Federal University of Goiás, Goiânia, GO, Brazil.
² Student of Medicine, Federal University of Goiás, Goiânia, GO, Brazil.

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INTRODUCTION

The fifth ventricle, also known as ventriculus terminalis (VT), is a term used to describe an ependymal-lined cavity of the caudal portion of the spinal cord. This cavity was first described by Stilling in 1859. The term “fifth ventricle” was only used in 1875, after observing that the walls of the cavity consisted of normal ependymal cell\(^1\). The fifth ventricle is located at the lower end of the conus medullaris, where the spinal cord is extended to the filum terminale just below the exit point of the last two coccygeal nerves\(^1\).

In a retrospective analysis of 418 pediatric spine magnetic resonance (MR) images, the isolated terminal ventricle was reported in 2.6%. All children with the fifth ventricle were younger than 5 years old and 72.7% younger than 1-year old\(^5\). Persistence of the ventriculus terminalis into adulthood is a rare entity, particularly in the absence of other structural malformations of the central nervous system\(^6\).

The imaging findings consist of intramedullary ovoid cystic lesion at the level of the conus medullaris\(^6\). Clinical manifestations vary, and conservative management is typically favored over surgery unless significant associated symptoms are present\(^7-9\).

We present a case report and literature review of a rare case of a child with symptomatic ventriculus terminalis and aim to emphasize the importance of including the fifth ventricle in differential diagnosis of the spinal cord lesions to ensure proper treatment.

METHODS

The reported patient was diagnosed and treated by the Division of Neurosurgery, Department of Surgery, Medical School, Federal University of Goiás (HC-UFG) in 2018.

The literature review was retrospective from 1991 to 2021 in Medline (PubMed) and Lilacs (via BVS) databases. The descriptors used were “persistent terminal ventricle” OR “Ventriculus terminalis” AND “children” and articles referring to case reports, case series and literature reviews on the subject were found and included.

CASE PRESENTATION

Male patient, 9-year-old, presenting episodes of lower limb paraparesis and sphincter dysfunction for 2 months. Physical examination revealed patellar hyperreflexia but no other neurological findings. He underwent a head and spinal MR. The spinal MR evidenced an intramedullary ovoid cystic lesion at the level of the conus medullaris, compatible with a terminal ventricle at the T11-T12 level (Figure 1 and Figure 2), and size of approximately 3.2 × 1.1 × 1.0 cm. Perilesional edema was not observed, and the head MR was normal.

Figure 1. Sagittal MR in T2 sequence showing cystic dilation (3.2 × 1.1 × 1.0 cm) at T11-T12 level. A. Preoperative image. B. Postoperative (2.7 × 0.5 × 0.4 cm) image.
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The surgery for fenestration of the V ventricle was performed in June 2018. The patient underwent a laminectomy at T11-T12, and then a midline myelotomy for cyst fenestration. Clear liquid similar to cerebrospinal fluid (CSF) was drained from the cyst. Postoperatively, a neurological improvement was evident during the first days after surgery. In follow-up, the patient presented good movement, associated with bladder and sensory normal functions. Four days after surgery, the patient was discharged without any neurological alterations.

However, on the 15th postoperative day, he presented with a CSF leak at the surgical wound without any phlogistic signs. The leak was surgically corrected on the same day and the patient was discharged 3 days later, without another complications.

Postoperative MR imaging revealed regression of the lesion, with approximately 2.7 × 0.5 × 0.4 cm.

**DISCUSSION**

Ventriculus terminalis has been described in children younger than 5-year-old as a normal asymptomatic developmental phenomenon. Rarely, the terminal ventricle persists until adulthood, when it becomes symptomatic, instead, our patient presented neurological abnormalities before that, within 9 years of age. Symptomatic ventriculus terminalis in children was only reported by one more article.

The mechanism that determines dilation of the ventriculus terminalis is still unclear, but several theories have been proposed to explain the origin of this pathological dilation. The formation of the *ventriculus terminalis*, *filum terminale* and part of the *conus medullaris* happens at the secondary neurulation. As the *filum terminale* is formed in a process that involves caudal regression and retrogressive differentiation (complete regression and disappearance of caudal tail by apoptosis), a malfunction on this phase could cause the deformation.

From a different perspective, Nassar et al. reported that cavitation may also be produced by vascular disturbance, inflammatory diseases, compression, or ischemic necrosis of the spinal cord. Sigal et al. postulated that physiological changes in CSF because of trauma may lead to abnormal closure and subsequent isolation of the ventriculus terminalis, precipitating enlargement of the VT into a cyst. Signs and symptoms from those triggers were not found in our patient report.

Besides the uncertainty of a definite pathogenesis, it is known that VT is detectable in the caudal neural tube by the 7th week after...
conception and on day 48 begins the retrogressive differentiation and subsequent regression of the structure.\textsuperscript{12} Despite the usual activation of this retrogressive process, this ependymal structure is often present in the conus medullaris in neonates and children, identifiable upon ultrasonography or MR\textsuperscript{14-17}. Spinal MRI imaging would usually show a cystic structure in the conus medullaris, not enhanced by contrast\textsuperscript{19}, compatible with our patient T11-T12 lesion (Figure 1 and Figure 2).

In summary, the fifth ventricle structure is seen during fetal development and in almost all infants. Before the age of 5, it is described as a normal developmental phenomenon which usually regresses; our patient's persistent ventriculus terminalis, a rare condition, which should be included in differential diagnosis of spinal cord lesions\textsuperscript{8,14-17}. Nevertheless, the few reported cases published until now persisted until adulthood without any symptoms related to the ventriculus terminalis\textsuperscript{8} and, to our knowledge, we present the second case in literature about symptomatic ventriculus terminalis on children and the first one with lower limb paresis on this age group.

Batista et al.\textsuperscript{19} presented a proposal for a clinical classification of the patients with cystic lesion of the ventriculus terminalis. The patients were divided into three groups based on clinical presentation classified as: Type I, nonspecific neurological, including low-back pain, sciatica, and inferior limb pain; Type II, presence of focal neurological deficit, including gait disturbances, paresis, sensory disturbances, altered deep or muscular atrophy (no complaints or findings of sphincter dysfunction); and Type III, presence of sphincter dysfunctions such as bowel and/or bladder dysfunction detected clinically, not explained by ultrasonography studies of the bladder or by urodynamic examination\textsuperscript{19}.

Although Batista et al.\textsuperscript{19} classification was designed for symptomatic adults, our patient was classified as a Type III cystic dilation of the ventriculus terminalis due to the bladder dysfunction, with concomitantly Type II symptoms, which correspond to 18 from 106 patients reviewed by Zeinali et al.\textsuperscript{9}.

The treatment of cystic lesions of the ventriculus terminalis is still controversial, as there are no high-quality, large sample prospective studies, which are limited by the rarity of the disease.\textsuperscript{20} Nevertheless, laminectomy followed by cyst fenestration is the most described procedure in literature. Batista et al.\textsuperscript{19} argued that Type I patients with nonspecific complaints are probably best treated conservatively because it avoids unnecessary procedures that would worsen the clinical state but affirmed that Type II and III patients can be successfully treated by surgical fenestration. At the same time Ganau et al.\textsuperscript{4} suggested that every patient, including those with Type I symptoms, improved immediately after surgery. Therefore, according to the current recommendations, our Type III ventriculus terminalis patient would be included in the surgically approached group.

In a small cohort, Fletcher-Sanderson et al.\textsuperscript{21} followed-up 13 surgical fenestrations and one conservative treatment. Two patients received cyst-subarachnoid shunt, and both did not present postoperative cyst recurrence, while three from the remaining eleven surgical patients recurred partially with the cystic dilation and the symptoms\textsuperscript{21}. Furthermore, they concluded that Type III patients, like our patient, should be approached by surgical fenestration to relieve symptoms and prevent acute cauda equina syndrome\textsuperscript{21}. Beside our case report, we did not identify any further description of a postoperative CSF leak from the surgical site. The patient remains asymptomatic after the fistula correction with no signs of relapse on control imaging.

Future studies should gather more patients to evaluate more accurate results to cyst fenestration surgery. Also, they should address cyst-subarachnoid shunt and the minimally invasive procedure called percutaneous aspiration using real time MR, which has reported good results until now\textsuperscript{22}.

We present a case report and literature review of a 9-year-old patient with episodes of paresis and bladder dysfunction due to a fifth ventricle at the level of T11-T12 that underwent a surgical resection with good neurological outcomes, without recurrence. Once again, even the symptomatic cases of ventriculus terminalis are difficult to be diagnosed due to their nonspecific symptoms, and spine MR is often capable of defining the cystic dilation.

We believe that this article was able to draw attention to a rare case of a symptomatic ventriculus terminalis, emphasize the importance of including the fifth ventricle in differential diagnosis of spinal cord lesions and to point out the gaps in the literature about the management of cystic lesions of the ventriculus terminalis.

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CONCLUSION

We present a case report and literature review of a 9-year-old patient with episodes of paresis and bladder dysfunction due to a fifth ventricle at the level of T11-T12 that underwent a surgical resection with good neurological outcomes, without recurrence. Once again, even the symptomatic cases of ventriculus terminalis are difficult to be diagnosed due to their nonspecific symptoms, and spine MR is often capable of defining the cystic dilation.

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CORRESPONDING AUTHOR

Lorimer Sandoval Carneiro, MD
Federal University of Goiás
Department of Surgery
Division of Neurosurgery
Goiânia, Goiás, Brazil
E-mail: lorimermd@gmail.com

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