Ventriculus terminalis

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The ventriculus terminalis (VT) is a small ependymal-lined cavity within the conus medullaris, and is as a result of canalization and retrogressive differentiation during embryonic development. The VT has been described as a normal developmental phenomenon in newborns and pediatrics, but is a rare pathology in adults, with only 31 cases reported to date ^{1) (2) (3) (4) (5)}.

Etiology

Remains unknown. Several hypotheses have been proposed for the development of VT. Some authors propose that cavitations might be associated with trauma, vascular disturbance, inflammatory, or compressive pathology of the spinal cord, and may interrupt communication between the VT and the central canal $^{6)$ 7).

In an initial report in 1859 by Stilling, the VT was identified as a ventricular structure enclosed by ependymal tissue and was called a terminal ventricle, an ependymal cyst, or the fifth ventricle. In the 7th week after conception, the VT is detectable in the caudal neural tube, regression, and regressive differentiation begins on day 48. Though the process of canalization and retrogressive differentiation may normally activate, this ependymal structure is often present in the conus medullaris, which becomes identifiable in neonates and children upon ultrasonography or MR imaging ^{8) 9) 10) 11)}.

In a postmortem study by Kernohan et al. ¹²⁾ the VT was reported to be a "true ventricle" and communicated with neither the subarachnoid space nor the central canal of the spinal cord. Coleman et al. ¹³⁾ reported that the dilated VT was found in 2.6% of pediatric patients (less than 5 years) without related symptoms. However, cystic dilatation of the VT in children may be associated with congenital anomalies, such as tethered cord syndrome, Chiari type I malformation, lipomyelomeningoceles, and lumbosacral lipoma ^{14) 15) 16)}.

Infrequently, the isolated dilatation of the VT is detected in the elderly, combined with clinical symptoms. Nassar et al. ¹⁷⁾ remarked that intramedullary cystic lesion of the conus medullaris may be produced by trauma, hemorrhage, compressive pathology, or vascular impairment. The maximal diameter of the cystic VT in adults is reported to be larger than that of the pediatric VT, which concurred with the results of Suh et al. ¹⁸⁾.

Sigal et al. ¹⁹⁾ described another hypothesis of the adult VT, which results from a lack of communication between the VT and the central canal. On this basis, CSF hydrodynamics of the adult VT was evaluated using specific MR imaging techniques. Recently ECG-gated SPAMM-MR imaging has been utilized to assess detailed motion and direction of CSF within the spinal cord ²⁰⁾. Wayte and Redpath ²¹⁾ reported visualization of pulsatile CSF by showing shift in tagging bands on the cardiac-gated cine images of SPAMM. In support, a few authors reported the evaluation of CSF hydrodynamics using the SPAMM technique in syringomyelia, cervical stenosis and Chiari malformation, concluding that SPAMM-MR imaging was useful in the quantification of spinal CSF flow ^{22) 23) 24) 25)}. In these reports, the presence of pulsatile motion in the syrinx represented communication with CSF space and the syrinx, as well as the redundant wall capacity of the spinal cord. In the study of Suh et al., ²⁶⁾ SPAMM-

MR imaging was performed in only 3 patients, which showed no shifts in the tagging band to the adjacent bands within the VT. According to these findings, they inferred that there was "no pulsatile motion within the cystic VT," supporting the previous theory as additional evidence.

Diagnosis

The characteristic feature of the VT in MR imaging is a cystic lesion of the distal central spinal cord canal without cord signal abnormality. However, in 3 patients who underwent surgical treatment (3/22, 13.6%), these lesions showed septations with cord edema, which were proven pathologically as an ependymal lining ²⁷⁾.



These unusual MR findings of the adult VT have been reported by Suh et al. ²⁸⁾ and the cause of these findings are still unclear, although compressive myelopathy and kyphosis may be associated with cord edema. It is very important to consider a differential diagnosis of spinal neoplasm if the cystic lesion of the conus medullaris has septation and edema.

Seo et al.²⁹⁾ reported on a non-enhancing intramedullary cystic lesion of the conus, which was pathologically confirmed as spinal astrocytoma and was not to be disregarded in the differential diagnosis of non-enhancing intramedullary lesions.

Treatment

The management of adult VT is controversial, as some prefer non-operative management with serial imaging, while others favor surgical maneuvers for relief of neurological symptoms ^{30) 31) 32) 33)}.

In the report of Suh et al., ³⁴⁾ surgical or endovascular procedures were performed in 4 patients with progressing neurological symptoms. After the procedure, 3 patients showed improved neurologic symptoms and back pain was stabilized in one. Most cases in previous reports had been treated with surgical maneuvers (89%, 16/18) and demonstrated improvement of their clinical symptoms.

In a review of the literature, a few authors reported on the postoperative clinical outcomes of the VT, which were evaluated according to their clinical presentation; patients with nonspecific neurologic

symptoms were managed conservatively, whereas patients with focal neurological deficits or sphincter disturbance underwent surgical management ³⁵⁾.

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