Cervical Myelopathy Caused by Engorgement of the Epidural Venous Plexus due to Cerebrospinal Fluid Overdrainage: Case Report and Review of the Literature

Mielopatia cervical causada por ingurgitamento do plexo venoso epidural devido à hiperdrenagem liquórica: relato de caso e revisão da literatura

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Abstract

This is a case report of a 33-year-old woman with cervical myelopathy caused by an enlargement of the cervical venous plexus, after she was submitted to a ventriculoperitoneal (VP) shunt that evolved to overdrainage.

Magnetic Resonance Imaging (MRI) revealed an epidural venous enlargement within the spinal channel, with a 50% narrowing from C2 to C5, and spinal cord compression. A shunt revision was performed using a programmable drainage system, and a second MRI revealed the absence of the venous enlargement, resulting in cervical spinal cord decompression and remission of neurological symptoms.

Compressive myelopathy consequent to the enlargement of the epidural venous plexus related to the overdrainage of the ventriculoperitoneal shunt system without typical signs of intracranial hypotension may result in misleading etiological diagnoses. Acknowledging this disorder is important to distinguish it from neoplastic processes or hematomas, for which surgical intervention may be needed.

Keywords
► overdrainage
► spinal dural enhancement
► ventriculoperitoneal shunt
► cervical myelopathy
► vertebral venous system
► cranio-cervical junction

Resumo

Este é um relato de caso de uma paciente de 33 anos de idade que apresentou mielopatia cervical devido a alargamento do plexo venoso cervical, após ser submetida a uma derivação ventriculoperitoneal que evoluiu com hiperdrenagem.
Introduction

The typical clinical manifestations of cerebrospinal fluid (CSF) overdrainage include orthostatic headache, nausea, vomiting, neck pain, diplopia and blurred vision.\(^1\) Brain magnetic resonance imaging (MRI) findings related to spontaneous intracranial hypotension (SIH) are rather variable: diffuse pachymeningeal enhancement associated with pituitary enlargement, optic chiasm flattening, increased anterioposterior diameter of the brainstem, and engorged cerebral venous sinuses may be encountered. Spinal MRI revealing an extra-arachnoid fluid collection, extradural extravasation of fluids, meningeal diverticula, diffuse pachymeningeal enhancement, and enlargement of the spinal epidural venous plexus are not uncommon,\(^2\) but compressive cervical myelopathy related to the epidural venous plexus engorgement caused by fluid hypotension is extremely rare.\(^3\)\(^-\)\(^11\)

This paper reports the case of a patient who presented a progressive cervical myelopathy related to an epidural venous plexus engorgement caused by shunt overdrainage. Since an early diagnosis was never reached, this case report covers the primary findings and pitfalls before the primary etiology was revealed.

Case Report

Case history: A 33-year-old woman was admitted to our service presenting a two year history of clinical cognitive progressive deficit, associated with visual alterations, episodes of loss of strength in the upper right limb, weakness in the lower limbs, and urinary retention. Both brain MRI and computed tomography (CT) exams revealed a communicating hydrocephalus associated with cortical atrophy (\(\text{Fig. 1}\)). Routine blood tests detected anemia, under only the hematological evaluation, thus the apparent cause of the anemia was not encountered.

The patient was submitted to a ventriculoperitoneal (VP) shunt with a high-pressure system. She developed a headache, which improved in repose, and control CT scans showed a well-positioned valve with reduction of the ventricular cavity.

Approximately two years after the VP shunt implant was done, neck pain, headache and visual deficit worsened, accompanied by quadriaparesis and paresthesia.

A cervical MRI evidenced the presence of an anterior cervical mass leading to significant spinal cord compression (\(\text{Fig. 2}\)), and a brain MRI showed the absence of...
hydrocephalus. Angioresonance showed a significant enlargement of the epidural venous plexus (► Fig. 3). The VP shunt overdrainage hypothesis had not been considered until then.

In another service, the patient was submitted to the replacement of the shunt system by a programmable pressure valve, and evolved to a progressive improvement in the neurological symptoms.

The control cervical MRI at one year revealed the complete absence of the cervical epidural mass (► Fig. 4).

Discussion

Venous drainage at the craniocervical junction flows through the internal jugular vein and vertebral veins with contribution of their associated anastomoses. The anterior and posterior condylar veins form the plexus of the vertebral vein, but this may be modified by the presence of emissary anastomoses of the mastoid, with an increased possibility of drainage at this level. The veins of the junction are formed by the confluence of the emissary and posterior cephalic veins with the superior epidural venous system.12

The contribution of the internal jugular veins for cerebral drainage is variable among individuals, with a possible venous flow ranging all the way from 6 to 72%.13 The epidural cervical venous system is located in the anterolateral portion of the spinal channel, and it may function as an alternative to the cranial drainage system, possibly affecting the intracranial pressure in some cases. It is equivalent to an independent accessory venous drainage system of the intracranial compartment.

The vertebral venous system may be divided into anterior and posterior sections, respectively consisting of the intraspinal epidural venous plexus and the external posterior paravertebral

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**Fig. 2** MRI images of the cervical spine two years after the VP shunt. T1-weighted (A), T2-weighted (B) and gadolinium-enhanced T1-weighted (C, D) demonstrating spinal cord compression at C2-C5.
Functional variations may occur depending on the flexion or extension and position of patient’s head.

Intracranial hypotension syndrome is characterized by postural headache and low fluid pressure. It may be classified into five types: primary or spontaneous; post lumbar puncture; cranial post-trauma; post-craniotomy; and severe depletion of volume. Ventriculoperitoneal shunt overdrainage is also thought to be another cause of intracranial hypotension syndrome.

Schaltenbrand\textsuperscript{16,17} introduced the concept of SIH and proposed three mechanisms to explain its pathophysiology: 1) decreased CSF production; 2) CSF hyperabsorption; or 3) CSF leak.

Spinal manifestations of SIH occur in nearly 6% of patients, and may involve all spinal levels with either myelopathy or radiculopathy. Diagnosis is not only difficult; in many cases, it is also late, given the infrequent spinal image evaluation, not counting the fact that nearly 20% of brain MRIs present as normal.\textsuperscript{18} Findings on brain MRI regarding SIH are variable. Diffuse pachymeningeal enhancement; pituitary enlargement; increased antero-posterior diameter of brainstem; subdural fluid collection; and engorged cerebral venous sinuses may be observed. Extra-arachnoid fluid collection, extradural extravasation of fluid, meningeal diverticula, diffuse pachymeningeal enhancement, and engorgement of the spinal epidural venous plexus may be noted in the spinal MRI.\textsuperscript{2}

The diffuse dural enhancement is considered to be the result of venous dilation caused by low CSF volume. According to what Monro-Kellie advocates, the intracranial volume system.\textsuperscript{14}
of CSF is inversely related to the cerebral blood volume. For
this reason, in SIH cases, a decreased CSF volume leads to an
increase in the dural venous blood volume and consequent
venous enlargement.19 This mechanism results in dural
thickening and enhancement. The Monro doctrine may be
applied to the vertebral channel, as both cavities, cranial and
spinal, are closed systems.8,20

Förderreuther et al were able to demonstrate through
cervical MRI the cervical epidural veins’ dilation during
orthostatic headache crises, with subsequent improvement
and remission of those signs once the pain subsided.20 Ciceri
showed evidence of vein dilation in Hirayama myelopathy
cases.21 The epidural venous plexus engorgement has been
described as another cause of radicular compression,22–24
and SIH was also shown to be a cause of dilation of the
epidural venous plexus.1

In an extensive review of the literature, not more than
10 related cases have been found – this present case
included – where a cervical venous plexus engorgement
results from shunt overdrainage (=Table 1). In 1998,
Miyazaki et al were the first to describe the clinical
manifestation of myelopathy in the overdrainage of a VP
shunt system in a patient with a low-pressure shunt after
subarachnoid hemorrhage.8 In the cases described, the
time interval to the onset of overdrainage symptoms after
shunt placement was quite variable. An interesting observa-

tion regards the initial manifestations of myelopathy,
which varied from 4 months to 10 years.3

Also noteworthy is the fact that no cases described in the
literature have presented the classic symptoms of CSF over-


drainage. The majority of patients have presented with
motor deficit clinical symptoms (9 cases), and only 1 case
presented with neck and back pain.6 The present case
initially presented with headache, which improved in re-
pose, and with neck pain after light trauma, further evolving
into the known fluid hypotension and compressive cervical
myelopathy symptoms.

Surgical treatment was needed in eight cases. Shunt
system replacement was the most common occurrence,
and clinical improvement was observed in all cases after
surgery. There was one case in which treatment was
refused,11 and another patient who only underwent clinical
follow-up.6 Complications such as the above mentioned
illustrate a rare clinical presentation of VP shunt failure,
reinforcing that adequate knowledge of this pathology is
paramount to its early identification.

Table 1 Cases described in the literature and results

<table>
<thead>
<tr>
<th>Literature</th>
<th>Sex/Age</th>
<th>Pathology</th>
<th>Symptoms/ time after VP shunt</th>
<th>Radiological findings</th>
<th>Treatment</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Miyazaki et al8</td>
<td>M/53</td>
<td>SAH</td>
<td>Quadriaparesis/ 4 years</td>
<td>Spinal cord compression CCj-C3</td>
<td>Ligation of VP shunt</td>
<td>Improvement</td>
</tr>
<tr>
<td>Matsumoto et al7</td>
<td>M/67</td>
<td>Pineal tumor</td>
<td>Paraparesis/ 2 years</td>
<td>Cranial diffuse meningeal enhancement</td>
<td>Removal of the shunt</td>
<td>Improvement</td>
</tr>
<tr>
<td>Wingerchuk et al11</td>
<td>F/72</td>
<td>Posterior fossa meningioma</td>
<td>Quadriaparesis/ 6 years</td>
<td>Cervical enhancement</td>
<td>Refused treatment</td>
<td>Unchanged</td>
</tr>
<tr>
<td>Liu et al5</td>
<td>F/18</td>
<td>Porencephalic cyst</td>
<td>Quadriaparesis/ 4 months</td>
<td>Engorgement of the epidural veins</td>
<td>Change to programmable shunt</td>
<td>Improvement</td>
</tr>
<tr>
<td>Humphries et al4</td>
<td>F/33</td>
<td>Dandy-Walker</td>
<td>Paraparesis/ 2 years</td>
<td>Engorgement of the epidural C2-C5</td>
<td>Change revision</td>
<td>Improvement</td>
</tr>
<tr>
<td>Wolfe et al10</td>
<td>M/17</td>
<td>Tumor cyst</td>
<td>Quadriaparesis/ 5 years</td>
<td>Dilated epidural venous plexus</td>
<td>Change to programmable shunt</td>
<td>Improvement</td>
</tr>
<tr>
<td>Martínez-Lage et al9</td>
<td>F/20</td>
<td>Communicating hydrocephalus</td>
<td>Neck and back pain/ 4 months</td>
<td>Lesion mimicking cervical epidural hematoma C1-C5</td>
<td>No surgical treatment</td>
<td>Improvement</td>
</tr>
<tr>
<td>Ulrich et al9</td>
<td>F/17</td>
<td>Obstructive hydrocephalus</td>
<td>Paraparesis/ 4 years</td>
<td>Engorgement of the cervical epidural venous plexus C2-C3</td>
<td>Change to anti-siphon device, valve pressure increased</td>
<td>Improvement</td>
</tr>
<tr>
<td>Cardoso et al3</td>
<td>M/32</td>
<td>Communicating hydrocephalus</td>
<td>Quadriaparesis/ 10 years</td>
<td>Engorgement of the cervical venous plexus CCj and intracranial</td>
<td>Change to programmable shunt</td>
<td>Improvement</td>
</tr>
<tr>
<td>Dantas et al (present study)</td>
<td>F/33</td>
<td>Communicating hydrocephalus</td>
<td>Quadriaparesis/ 2 years</td>
<td>Spinal cord compression by engorgement of the cervical venous plexus C2-C5</td>
<td>Change to programmable shunt</td>
<td>Improvement</td>
</tr>
</tbody>
</table>

Abbreviations: CCj, craniocervical junction; F, female; M, male; SAH, spontaneous subarachnoid hemorrhage; VP, ventriculoperitoneal.
Cervical Myelopathy due to Cerebrospinal Fluid Overdrainage

Dantas et al.

Conclusion

This case is an interesting presentation of clinical failure of the shunt, with overdrainage involving the cervical venous plexus and venous cerebral drainage. The engorgement of the cervical venous plexus should always be kept in mind during differential diagnoses of myelopathy in patients with shunt and clinical signs of overdrainage.

Disclosure

The authors report no conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper.

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