Spinal dural arteriovenous fistula presenting with paraplegia following lumbar puncture

Case report

GUUS KOERTS, M.D., VINCENT VANTHUYNE, M.D., MAXIME DELAVALLEE, M.D., HERBERT ROOIJAKERS, M.D., AND CHRISTIAN RAFTOPOULOS, M.D., PH.D.

Department of Neurosurgery, Cliniques Universitaires St-Luc, Brussels, Belgium

Spinal dural arteriovenous fistulas are rare lesions with an annual incidence of 1 per 100,000 population. In patients with this disease, an abnormal vascular dural shunt exists between a dural branch of a segmental artery and a subdural radicular vein that drains the perimedullary venous system, leading to venous hypertension and secondary congestive myelopathy. Generally, patients present with progressive paraparesis, urinary disturbances, and gait ataxia. In this report the authors describe a 61-year-old woman with a spinal dural arteriovenous fistula who developed an acute paraplegia after a nontraumatic lumbar puncture. The possible underlying mechanisms and treatment options are discussed.

(Please refer to the original source for the complete text.)

Abbreviations used in this paper: AVF = arteriovenous fistula; LP = lumbar puncture; MRC = Medical Research Council; SDAVF = spinal dural AVF.

Case Report

History and Examination. This 61-year-old woman developed progressive walking problems in January 2006. Five months later she was only able to walk with the aid of 2 crutches, and she also reported pain at the level of the thoracic spine. Initial clinical examination and MRI sequences were performed in another clinic. Their neurological evaluation revealed a proximal paraparesis (MRC Grade 4/5) and an ataxic gait. The MRI studies (Fig. 1) showed intramedullary pencil-like T2 hypersignal and dilated perimedullary veins. Based on the clinical findings and MRI results, a differential diagnosis of myelitis, intramedullary tumor, or ischemic lesions secondary to an AVF was made. The patient subsequently underwent an LP in the sitting position and with anteflexion. The LP was immediately followed by paraplegia, which was observed by her neurologist. Two days later, on arrival at our institution, clinical examination confirmed paraplegia (Aminoff-Logue Grade 5 and McCormick Grade IV), a sensory deficit below T-10, and urinary retention. Control MRI studies excluded hematoma. Spinal angiography (Fig. 2) demonstrated the presence of an SDAVF depending on the left L-2 radicular artery. Endovascular occlusion was not possible because the L-2 radicular artery, actually the Adamkiewicz artery, was also filling the anterior spinal artery.

Operation. A laminectomy at the level of L2–3 was performed. After opening the dura mater we observed a...
dilated perimedullary venous plexus. The CSF was clear (no subarachnoid hemorrhage) and circulated normally. A dilated arterialized vein was identified going to the left L2–3 foramen. Directly adjacent to this vein, a small radicular artery and the artery of Adamkiewicz were seen. We considered that the SDAVF was in the dural sleeve more laterally, and thus was not visible. After clip application and coagulation, the draining vein was sectioned intradurally at the level of L2–3.

Postoperative Course. The patient improved in the early postoperative phase to a paraparesis graded 2/5 (MRC scale). Her hypesthesia regressed as well. On the 11th postoperative day she was transferred to a rehabilitation center. Three months later an almost full motor recovery was present, allowing the patient to walk with a stick, although her gait remained ataxic. Control MRI showed a regression of the intramedullary hypersignal. Control spinal angiography confirmed complete exclusion of the SDAVF. At the latest follow-up (53 months) the patient was able to walk with the aid of a cane. Although there was a regression of the intramedullary hypersignal, she suffers from severe neuropathic pain in her legs requiring medical treatment.

Discussion

Our patient with an SDAVF developed an acute temporary paraplegia following LP. Although acute paraplegia has been reported due to compressive hematoma or CSF obstruction following LP, control spinal MRI excluded these mechanisms in our patient (Table 1).2,12,30 An SDAVF is a rare lesion in which an abnormal vascular shunt exists between a dural branch of a segmental artery and a radicular vein that drains the perimedullary venous system.6,19,24,33 The pathophysiological mechanism of venous hypertension and secondary congestive myelopathy has been confirmed by pathological, hemodynamic, and radiological studies.3,10,11,13,17,19,24 Acute deterioration after epidural injections in patients with an SDAVF has been reported.11,29 Injection of a volume in the epidural space will increase the epidural pressure and subsequently decrease the drainage from the arterialized spinal cord venous plexus to the extradural veins.11 Injection of a small volume in these patients may be enough to exacerbate venous hypertension in the spinal cord. In our patient, however, no volume was injected. The occurrence of activity-exacerbated symptoms has been reported.3,20,26 Valsalva-like maneuvers will further increase venous pressure and compromise spinal cord venous drainage.20 Lumbar puncture is often done with the patient in flexion and with abdominal compression equivalent to a passive Valsalva maneuver.

The Monro-Kellie hypothesis was initially applied to the cranial contents, but MRI studies in patients with intracranial hypotension or CSF volume depletion have
demonstrated engorgement of cerebral venous sinuses and spinal venous plexus. Decreases in CSF pressure (spontaneous or iatrogenic [LP]) can be accompanied by engorgement of epidural and intradural veins and increased medullary ischemia.

As in our case, 3 other patients have presented with neurological deterioration after LP or myelography (Table 1). A possible mechanism of acute deterioration in patients with SDAVF and LP could be that a sudden decrease of CSF pressure will lead to dilation of intradural and extradural veins. Together with the Valsalva-like maneuver during LP, this could be enough to explain an acute ischemic aggravation and the acute symptomatology of these already vascularly compromised patients. In patients with clinical symptoms and typical MRI appearance (intramedullary pencil-like T2 hypersignal and dilated perimedullary veins) of SDAVF, LP should be avoided and spinal angiography performed to confirm the diagnosis. To our knowledge there is no indication for acute deterioration in patients with SDAVF receiving steroids. On the contrary, acute neurological deterioration after LP or myelography (Table 1) has been reported.

Steroids produce hydrostatic redistribution of intracranial, intraspinal, and extradural veins. Together with the Valsalva-like maneuver during LP, this could be enough to explain an acute deterioration in patients with SDAVF receiving steroids. On the contrary, acute neurological deterioration after LP or myelography (Table 1) has been reported even in patients who have been treated with steroids. Nevertheless, motor improvement has been reported even in patients who have been paraplegic from 3 months to more than 6 years.

Table 1: Acute paraplegia and paraparesis in patients with SDAVF or arteriovenous malformation after LP

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Spinal Level</th>
<th>Clinical Presentation Post-LP</th>
<th>Clinical Post-LP</th>
<th>Postpost Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Roulet et al., 1988</td>
<td>T-6</td>
<td>paraplegic</td>
<td>retention</td>
<td>paraplegic</td>
</tr>
<tr>
<td>T-7</td>
<td>severe paraparesis</td>
<td>retention</td>
<td>walking</td>
<td>normal</td>
</tr>
<tr>
<td>Awad &amp; Barnett, 1990†</td>
<td>T7–8</td>
<td>paraplegic</td>
<td>retention</td>
<td>walking</td>
</tr>
<tr>
<td>Aloui-Kasbi et al., 2004‡</td>
<td>NR</td>
<td>paraplegic</td>
<td>incontinence</td>
<td>paraplegic</td>
</tr>
<tr>
<td>present study</td>
<td>L2–3</td>
<td>paraplegic</td>
<td>retention</td>
<td>walking</td>
</tr>
</tbody>
</table>

* NR = not reported.
† Spinal arteriovenous malformation.
‡ Traumatic LP.

References
33. Thron A: [Spinal dural arteriovenous fistulas.] Radiologe 41:955–960, 2001 (Ger)

Please include this information when citing this paper: published online May 3, 2013; DOI: 10.3171/2013.3.SPINE12888.
Address correspondence to: Guus Koerts, M.D., Department of Neurosurgery, Cliniques Universitaires St-Luc, Avenue Hippocrate 10, Brussels 1200, Belgium. email: guus.koerts@uclouvain.be.