Tethered cord due to caudal lipomeningocele associated with a lumbar dural arteriovenous fistula

Case report

SANDIP B. MAVANI, M.C.H., AND TRIMURTI D. NADKARNI, M.C.H.

Department of Neurosurgery, King Edward Memorial Hospital, Seth G.S. Medical College, Parel, Mumbai, India

A 29-year-old man presented with progressive paraparesis associated with sensory impairment in both lower limbs for the past 2 years. He was experiencing the sensation of incomplete urinary evacuation. The patient had undergone an earlier operation for a lumbar lipomeningocele at birth. Magnetic resonance images of the lumbosacral spine showed a low-lying conus medullaris adherent to a caudal lipoma. There was a leash of abnormal vascular channels in the adjacent subarachnoid space. The patient underwent spinal angiography that revealed a dural arteriovenous fistula (AVF) principally fed by the left fourth lumbar (L-4) radicular branch. At surgery the cord was detethered by disconnection of the sacral lipoma. The dural fistula was obliterated by occlusion of the L-4 radicular feeder close to the nidus of the fistula. Postoperatively, the patient experienced an immediate relief of sensation of tightness in both lower limbs. There was a gradual improvement of power and sensation at the 6-month follow-up examination. According to the authors’ literature search, the present case is a unique report of a rare association of spinal cord tethering due to a caudal lipoma associated with a lumbar dural AVF. The present report discusses the etiopathology, presentation, and management of this case.

**Key Words** • tethered cord • low-lying conus medullaris • spinal lipoma • dural arteriovenous fistula • spinal angiogram • congenital

Abbreviations used in this paper: AVF = arteriovenous fistula; AVM = arteriovenous malformation.
line incision was noted at the lumbosacral spine. The fifth lumbar (L-5) spinous process was not palpable.

Magnetic resonance imaging of the lumbosacral spine showed a low-lying conus medullaris adherent to a caudal lipoma (Fig. 1A). A large subcutaneous lumbosacral lipoma was noted. Axial T2-weighted MR images showed the caudal lipoma exiting through spina bifida at the L-5 level (Fig. 1B). It was obvious from these images that, at the previous surgery during childhood, the subcutaneous lipoma had been superficially disconnected and detethering had not been achieved. The conus was noted at the L4–5 level and was associated with multiple flow voids in the adjacent dorsal subarachnoid space. This leash of abnormal venous channels was highly suggestive of a spinal vascular malformation. Spinal angiography showed a dural AVF principally fed by the left L-4 radicular artery (Fig. 1C). Urodynamic studies documented a neurogenic bladder of lower motor neuron type.

Surgery and Postoperative Course. At surgery the cord was detethered by disconnection of the sacral lipoma. The dural fistula was obliterated by occlusion of the left L-4 radicular feeder close to the nidus of the fistula. There was an immediate change in the color of the draining arterialized venous channels. The turgid vessels collapsed. The nidus of the fistula was excised. Postoperatively, the patient experienced symptomatic relief of tightness in both lower limbs. There was a gradual improvement of power and sensation as well as bladder function at the 6-month follow-up evaluation.

Discussion

Spinal lipomas are rare, with an overall incidence approaching 1% of all spinal axis tumors.4,6 In 1982, Chapman classified 3 anatomical variants of lipomeningocele according to the relationship of the lipoma–spinal cord interface: dorsal, caudal, and transitional.1 According to Pang, a fourth classification of lipomeningocele not previously described in the literature is the chaotic form. This type of lipomeningocele contains “unruly internal structures that do not obey the embryologic logic of either dorsal or transitional lipoma.”11

Fig. 1. Preoperative images of the patient. A: Sagittal T2-weighted MR image of the lumbosacral spine showing a low-lying conus medullaris. The lower dorsal cord demonstrates hyperintense cord signal intensity suggestive of venous hypertension. Multiple flow voids are noted in the subarachnoid space. A large lumbosacral subcutaneous lipoma is noted. B: Axial T2-weighted MR image demonstrating spina bifida of the L-5 vertebra. The conus is tethered to the subcutaneous lipoma. C: Spinal angiogram shows a dural fistula in the lumbar region fed principally by the left L-4 radicular artery. The characteristic corkscrew venous channels are noted.
Tethered cord and lumbar dural AVF

Spinal AVMs constitute 3.4% to 11.5% of spinal cord pathological lesions. It is now generally recognized that the spinal vascular abnormalities are not a single entity but consist of several biologically distinct forms, and 4 major types of spinal vascular abnormalities or spinal axis AVMs are now recognized: dural AVFs (Type I), intramedullary AVMs with or without extramedullary extension (Types II and III), and perimedullary AVFs (Type IV). The present patient had a caudal lipomeningocele associated with a dural AVF. The spinal vascular lesion of our patient was of the Type I variety. Recently, Spetzler et al. introduced a modified classification system to clarify issues raised by the existing nomenclature. Using the modified classification, our current case would be termed an intradural dorsal AVF.

Unlike intradural AVMs, dural AVFs have a strong male predilection and develop in the latter half of life. Dural AVFs have a strong tendency to occur in the lower thoracic and lumbar regions. Consequently, patients with spinal dural AVFs are unlikely to exhibit upper-extremity involvement and typically have an insidious onset of paraparesis or sphincter dysfunction. Low-back or radicular pain often precedes the gradual onset of the condition. In progressive myelopathy, Patients with dural AVFs frequently report worsening of symptoms during physical exertion (neurogenic claudication) or with certain changes in posture. It is interesting to note that the symptoms of a dural AVF mimic those of tethered cord syndrome, and it is clinically not possible to differentiate between the two. It is also not possible to suspect the occurrence of a dural AVF based on symptoms alone.

The paraparesis in the present patient could have been due to the stretching caused by the tethering of the cord, or related to the venous hypertension caused by the dural AVF, or both. In animals, Yamada and coworkers have shown that caudal traction on the cord results in impairment of oxidative metabolism and that the degree of impairment correlates with the severity of the neurological deficits. It was then postulated that such cord traction, as occurs in lipomeningocele cases, causes traction-induced hypoxia and stretching of the neuronal membrane with “loss of transmembane ion homeostasis and electrical activity depression.” A dural AVF results in venous hypertension. This venous congestion, with the subsequent decrease in spinal perfusion pressure, is widely accepted as the pathophysiology of the spinal cord ischemia that causes a deteriorating clinical course in these patients. Thus, in both tethered cord and dural AVFs, cord ischemia due to either spinal cord traction or venous stagnation results in neurological deterioration.

Dural AVFs are principally an acquired pathology. They have been known to occur in posttraumatic cases and postoperatively. However, according to Pia and Djindjian, spinal AVMs coexist with other congenital abnormalities—including cutaneous angiomata, vertebral anomalies, vertebral hemangioma, Rendu-Osler-Weber syndrome, Cobb syndrome, and venous and lymphatic dysplasia—in as many as 25% of cases. Although spinal AVMs are often associated with other cutaneous and vascular abnormalities, the coexistence of spinal AVFs and lipomas is exceptional; only 6 cases have been reported in the literature (Table 1). Among these 6 cases, only 2 are related to an intrathecal lipoma. In the first case reported by Djindjian et al., a lipoma of the filum terminale was associated with a sacral dural AVF in a 53-year-old man. Rice and Jelsma have described a combined intramedullary AVM and lipoma at the cervicothoracic level in a 23-year-old woman. The coexistence of the two pathologies in these cases appears to be congenital. It is possible that the lipoma itself and its tethering effect further impairs the venous drainage and leads to increased symptomatology of the fistula. Regardless of the etiology, it is important to note that dural AVFs may occur within the setting of a sacral lipoma and tethered cord syndrome and should be considered in the diagnostic evaluation of these lesions.

Table 1 presents all cases of spinal lipomas associated with AVMs available in the literature, including the present report. Although all cases represent a similar group of patients, there are differences in the patients based on location of the spinal tethering and the nature of associated arteriovenous vascular anomalies. The present case is unique and rare because the occurrence of both the lipoma and dural AVF are in the lumbar region. In the other reported cases, the lipomas were situated in anatomically different areas of the spine (cervical, thoracic, and sacral). In the studies by König et al. and Kendall and Logue, the patients harbored lipomeningoceles, as was noted in our patient in the lumbar region. However, among these patients, the site and nature of the AVMs were different. In the study by König et al., the lumbar lipoma was associated with a thoracolumbar extraspinal dural AVF, and in the study by Kendall and Logue, the lumbar lipoma was noted with a sacral AVM. Hence, the present case is obviously anatomically and pathologically a different clinical entity.

The authors cannot make conclusions about the etiology, process of development, or association between the two different pathologies that coexisted in the present case (a lumbar dural AVF and a lipomeningocele); the pathologies could have originated either congenitally or could have been acquired in later life. The initial neuroimages, radiological reports, and details of the first surgery were unavailable due to the length of time that had passed between the procedures. However, the present imaging of the patient suggested that detethering had not been achieved at the first surgery, as the intradural lipoma and its subcutaneous extension were clearly observed in the axial image. Thus, it is clear that only the subcutaneous lipoma was addressed at the first surgery and that the initial repair was incomplete and no intradural procedure had been performed. Therefore, it remains unclear whether both of these lesions coexisted congenitally or if the long-term tethering has precipitated the subsequent formation of the dural AVF.

Spina bifida and cord tethering, as well as spinal AVMs, are both congenital in origin when they occur independently. In the available literature, only 7 cases (including the present case) of tethered cord have presented with associated spinal AVMs. Based on this observation, the authors conclude that long-term tethering is unlikely to precipitate spontaneous formation of a spinal AVM. Furthermore, considering the number of spinal lipomas...
TABLE 1: Review of the 7 cases of spinal lipomas associated with AVMs *

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Clinical Presentation</th>
<th>Site of Lipoma</th>
<th>Nature of Spinal AVM</th>
<th>Treatment Modality</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>present case</td>
<td>29, M</td>
<td>paraparesis w/ sensory impairment &amp; bladder involvement for 2 yrs</td>
<td>lumbar LMC</td>
<td>dural AVF supplied by Lt L-4 radicular artery</td>
<td>lipoma: complete detethering; dural AVF: resection</td>
<td>improved in motor power &amp; incontinence at 6-mo follow-up</td>
</tr>
<tr>
<td>Cheung et al., 2005</td>
<td>41, M</td>
<td>severe paraparesis &amp; urinary incontinence for 4 wks</td>
<td>sacral (S1–4) lipoma</td>
<td>pial AVF at S1–2</td>
<td>lipoma: partial resection; AVM: resection</td>
<td>after 18 mos patient ambulated using a brace; bladder incontinence not resolved</td>
</tr>
<tr>
<td>Lee et al., 2000</td>
<td>44, M</td>
<td>stiffness &amp; paresthesia in both lower limbs for 7 yrs</td>
<td>thoracolumbar LMC (T11–L1)</td>
<td>intramedullary AVM at T11–L1 supplied by T10–12 intercostal arteries &amp; L-1 lumbar artery</td>
<td>lipoma: partial resection; AVM: embolization</td>
<td>complete neurological recovery</td>
</tr>
<tr>
<td>König et al., 1999</td>
<td>50, M</td>
<td>paraplegia, sensory loss at T-10, bowel &amp; bladder dysfunction</td>
<td>lumbar LMC</td>
<td>thoracolumbar extradural AVM supplied by 3rd lumbar artery</td>
<td>lipoma: resection; AVM: resection</td>
<td>not available</td>
</tr>
<tr>
<td>Djindjian et al., 1989</td>
<td>53, M</td>
<td>paraparesis for 1 yr</td>
<td>filum terminale lipoma</td>
<td>sacral dural AVF supplied by lateral sacral artery</td>
<td>lipoma: complete resection; AVM: resection</td>
<td>unchanged at 6-mo follow-up</td>
</tr>
<tr>
<td>Rice &amp; Jelsma, 1986</td>
<td>23, F</td>
<td>spastic paraparesis for 4 wks</td>
<td>cervicothoracic (C7–T2) intramedullary lipoma</td>
<td>intramedullary AVM at T1–2 level supplied by the thyrocervical trunk</td>
<td>lipoma: partial resection; AVM: embolization</td>
<td>ambulatory at 6 mos w/ improved urinary incontinence</td>
</tr>
<tr>
<td>Kendall &amp; Logue, 1977</td>
<td>3, F</td>
<td>paraparesis, sensory deficit below T-6 &amp; urinary incontinence for 1 yr</td>
<td>LMC</td>
<td>sacral AVM supplied by subcostal artery</td>
<td>surgery</td>
<td>excellent recovery in motor strength; bladder control returned to normal</td>
</tr>
</tbody>
</table>

* LMC = lipomeningocele.

that are and have been detethered, the incidence of postoperative occurrence of a dural AVF is nearly non-existent. It is highly improbable that the dural AVF formed spontaneously after spinal cord detethering or that the previous surgery could have contributed to the development of the dural AVF. Based upon the above data of the present and reported cases, a congenital association appears to be strong and more probable.

Clinically, if a spinal AVM is suspected as a possible co-diagnosis with lipoma based on radiological information, selective spinal angiography is recommended before surgery to identify the type and location of a spinal AVM and its feeders. The diagnosis of the dual lesions may be missed if angiography is not performed. Cheung et al.² have reported on a patient who had a tethered cord associated with a dural AVF located at the S1–2 level. The AVF was overlooked at the first surgery and required obliteration 8 months later after the patient had undergone neurological deterioration.

Based on the current knowledge of spinal dural AVFs, our treatment strategy involves the disconnection of the AVF from the venous drainage to relieve venous pressure and restore adequate cord perfusion.⁶ Currently, these lesions can be managed either surgically or endovascularly. However, as noted in our case, when dual lesions coexist, both detethering and obliteration of the dural AVF can be performed microsurgically at the same operation.

The dual occurrence of lipomeningocele with spinal vascular formations needs to be borne in mind. Neuroradiology clearly provides evidence to the concurrent existence of these lesions. Spinal angiography clearly defines the anatomy of the spinal vascular malformation. Surgical treatment of both pathologies during the same operation can provide a complete cure and the best chance of neurological recovery.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: Nadkarni. Administrative/technical/material support: both authors. Study supervision: both authors.

References

2. Cheung AC, Kalkanis SN, Ogilvy CS: Paraplegia after tethered cord surgery: an uncommon combined anomaly of spinal
Tethered cord and lumbar dural AVF


Manuscript submitted July 18, 2013.
Accepted May 22, 2014.

Please include this information when citing this paper: published online June 27, 2014; DOI: 10.3171/2014.5.SPINE13670.

Address correspondence to: Trimurti D. Nadkarni, M.Ch., Department of Neurosurgery, King Edward Memorial Hospital, Seth G.S. Medical College, Parel, Mumbai 400012, India. email: tdnadkarni@hotmail.com.