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Hydromyelia Associated With Spinal Lipoma of the Conus

Case Report

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Study Design. A case report and literature review of the treatment of "noncommunicating" syringomyelia.

Objective. The aim of this report is to document the timing and the treatment of hydromyelia holocord after surgical treatment for both tethering and retethering of spinal lipoma.

Summary of Background Data. Syringomyelia associated with spinal lipoma presents a different pathogenesis and treatment in comparison to the "communicating" hydromyelia in the myelomeningocele. After the primary retethering operation performed in symptomatic patients, recurrent retethering can occur with an increase of the syringomyelia signs and symptoms.

Methods. Syringomyelia treated with a thin silastic tube passed from the syrinx to the subarachnoidal space for drainage and decompression. Prior operations were: (1) initial untethering at birth, (2) second untethering at 5 years of age, (3) posterior fossa and cervical decompression.

Results. Magnetic resonance imaging 6 months post shunt operation demonstrated decompression of the hydromyelia holocord and syringobulbia with improvement of motor function of the legs and improvement in sensory symptoms.

Conclusion. Usefulness of syrinx-subarachnoidal shunt is demonstrated in this case report after unsuccessful decompression and detethering. When the enlargement of the ependymal channel is greater than 50% of the spinal cord's diameter, neurologic, and urological symptoms are evident and the patient benefitted from cord untethering and syrinx drainage. (1) The terminal "noncommunicating" syringomyelia in lumbar sacral lipoma has been reported to be associated with retethering in spinal lipoma in the 25% of the cases. (2) The rise of distal syringomyelia isn't only linked to the kind of the spinal lipoma, but also to the difficulty to obtain the untethering and a smooth cerebrospinal fluid flow between the subarachnoidal space and the ependymal canal. (3) In patients with hydromyelia holocord greater than the 50% of the spinal cord's diameter a myelotomy and insert an ependymal channel/syrinx to the subarachnoidal space shunt can resolve of the syrinx. In this case, the enlargement of the ependymal channel in "noncommunicating" syringomyelia associated with lumbosacral lipoma is greater than 50% of the spinal cord's diameter; neurologic and urological symptoms occurred and the patient benefited from cord untethering and concurrent syrinx drainage.

Key words: occult spinal disraphisms, tethered spinal cord, hydromyelia. **Spine 2010;35:E1069–E1071**

The advent of the magnetic resonance imaging (MRI), in the last decades, gave the opportunity to distinguish 2 kinds of syringomyelia associated with spinal dysraphisms: "communicating" in the myelomeningocele, with Arnold Chiari malformation and hydrocephalus[1,2] which extends over the dorsal spine and "noncommunicating" in lumbar sacral lipoma that occurs adjacent to the distal spinal cord.[3] The terminal "noncommunicating" syringomyelia has been reported to be associated with retethering in spinal lipoma in the 25% of the cases. The authors describe the clinical/neuroradiologic course and the surgical approach in a child with a hydromyelia holocord and a syringobulbia after 10 months from the untethering performed at the age of 5. In literature, there are few articles[4] about the treatment of partial syrinx in patients with dysraphisms, but none describe children affected by syringomyelia holocord with the aim to deduce the pathologic mechanisms and the surgical treatment option other than direct detethering/resection of the lipoma.

Case Report

The infant, at birth, presented a subcutaneous lipoma in the lumbosacral region; MRI showed a L4–S2 dorsocaudal lipoma with the distal spinal cord at the L5–S1 level and slight enlargement of the ependymal channel rostral to the lipoma, but no Chiari malformation or hydrocephalus. The neurologic evaluation demonstrated motor weakness of the lower extremities, paresis of the feet and hypotrophy of the gastrocnemius muscles. The ultrasound of the bladder, pre/post void, and urodynamic studies showed a detrusor sphincter dyssynergia. The SEPs and EMG confirmed the damage of the spinal cord and nerve roots L4–S1. This infant's initial operation, 40 days after the birth, was a lumbar spinal lipoma repair with a L5 laminectomy, incomplete untethering for partial removal of the intradural lipoma and short sacral nerve roots. Five years later, the child was presented with low back and left leg pain, worsening weakness of the lower extremities, and gait deterioration. A spinal MRI showed a spinal cord tethered and minimal enlargement of the ependymal channel from L5 to T8 level. The urodynamic study confirmed a detrusor sphincter hyperreflexia. The patient then had a second untethering operation. Seven months after the second

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untethering operation the child presented with a rapid loss of the voice, hiccup, and worsening of the gait. A spinal MRI showed a hydromyelia holocord and a syringobulbia (Figure 1). The child had a third operation at this time of a sub occipital craniectomy and an upper cervical laminectomy, lysis of arachnoidal adhesions, drainage of the fourth ventricle, and duraplasty in attempt to relieve the syringobulbia. With the reappearance of hiccup and gait deterioration, after 40 days, a repeat new cerebral and spinal MRI demonstrated a worsening of the hydromyelia with an enlargement of the ependymal channel greater than 50% of the spinal cord's diameter. The patient subsequently underwent an intradural exploration of the lumbar sacral space with dissection of the subarachnoidal adhesions, to restore normal regional cerebrospinal fluid (CSF) flow patterns. During this operation a thin silastic tube was also passed 10-cm proximally in the syrinx, via a dorsal midline myelotomy immediately rostral to the lipoma. The distal end of the tube was left in the subarachnoidal space. The tube was secured to the spinal arachnoidal layer at the site of the myelotomy with 6–0 suture. The neurologic postoperative evaluation revealed an improvement in motor function of the legs, significant improvement in sensory symptoms, but unchanged fecal and urinary incontinence. MRI after 2 weeks demonstrated an initial reduction of the hydromyelia holocord and syringobulbia, with complete resolution 6 months after the operation (Figure 2). The clinical symptoms have remained stable for 38 months from the fourth operation which shunted the syrinx.

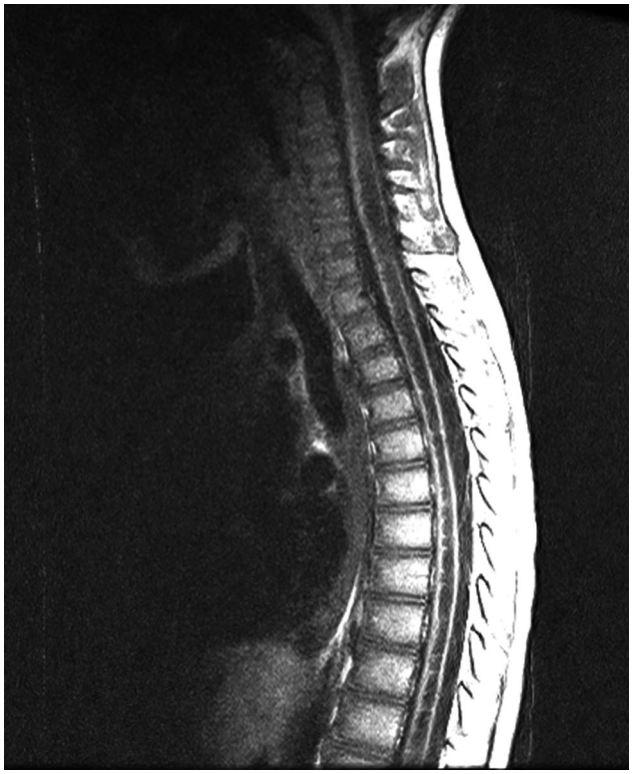

Figure 1. Pre-posterior fossa decompression sagittal T1-weighted MRI scan demonstrating a syringobulbia and dorsolumbar syringomielia.

■ Discussion

The syringomyelia in the spinal lipoma presents a different pathogenesis and treatments in comparison to the "communicating" hydromyelia in the myelomeningocele. The extension and the degree of enlargement of the terminal syrinx appeared to be correlated with a tethered cord syndrome. At presentation patients with a spinal lipoma and a pre-existent enlarged central channel, already present neurologic, urological, and orthopedic deficits. The primary operation performed in these symptomatic patients is often associated with new retethering and increase of the syringomyelia[5] that can suggest further surgical treatments. This progression and evolution of symptoms and radiologic imaging of syrinx size increase can be explained with an occlusion in the first years of life between ependyma-lined channels within lumbar lipoma and the ependymal channel,[6] already present at birth. The authors, in the prior lumbar sacral lipoma repair, recommend the lowest intact lumbar spinous process laminectomy and exposition of the dural sac; to perform a microsurgical intradural exploration and excision of the subarachnoidal adhesions, of all the fibro neural bands and of the lipoma that tether the spinal cord dorsally to the adjacent dural sac in order to avoid a CSF blockage between the arachnoidal space and the ependymal channel, identification, and section of the terminal filum.[7,8] We think that the rise of distal syringomyelia is not only linked to the kind of the spinal lipoma, but also to the difficulty to obtain the untethering and a smooth CSF flow between the subarachnoidal space and the ependymal canal. Although the treatment of the syringomyelia is not necessary in the great majority of cases, in our patient the syringomyelia holocord was linked to the distal lipoma, not to the obstruction of the outlets of the fourth ventricle. This fact was demonstrated because the syringomyelia was present before and grew after performing the posterior fossa decompression. In this case with syringobulbia and hydromyelia holocord greater than the 50% of the spinal cord's diameter demonstrated that the initial distal untethering and sub occipital craniectomy were ineffective managements in this patient. The reason behind the craniocervical decompression was to prevent a rapid development of bulbar symptoms into a possible fatal suffering of the brain stem. However, a fourth operation with a myelotomy and insertion of a shunt catheter between the ependymal channel/syrinx and the spinal subarachnoidal space obtained a resolution of the syrinx.[8,9] The current medical knowledge to explain the pathogenesis of the terminal syringomyelia[10,11] are recognized to have several limitations. More patient case reports with surgical outcome data are needed to more fully understand the mechanism of development, the timing and the ideal treatment of the syringomyelia in the spinal lipoma. The patients with lumbar lipoma and syringomyelia at birth need scheduled multidisciplinary evaluations to detect the onset of progressive neurologic deficits and poten-

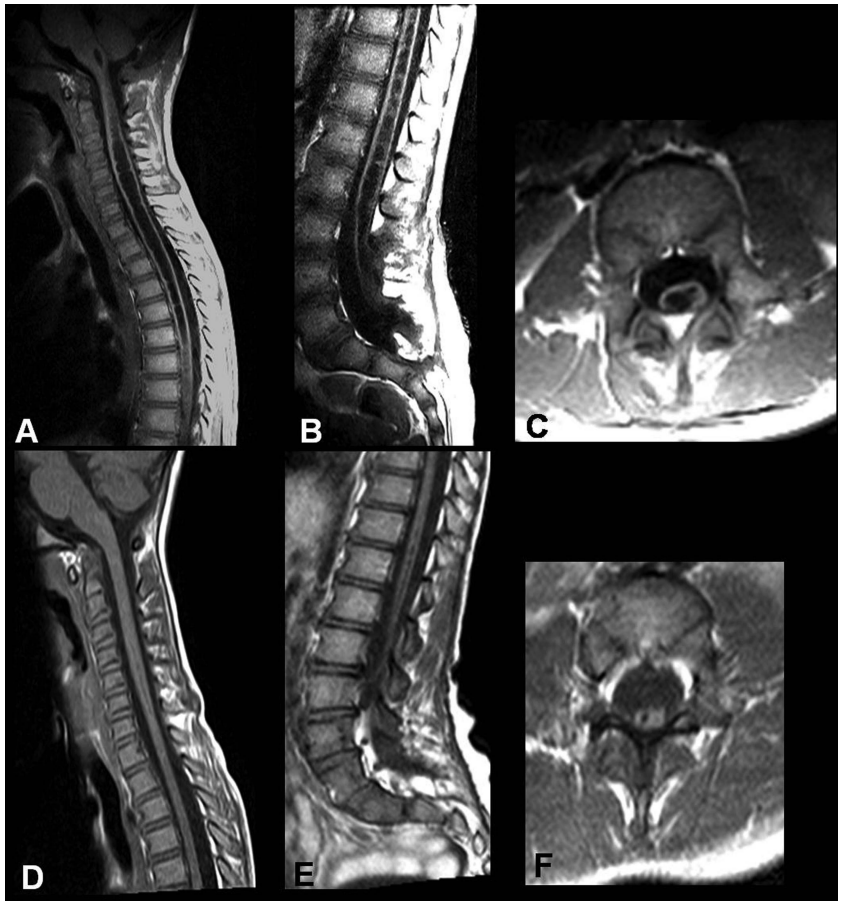

Figure 2. **A**, **B**, **C**, Preoperative sagittal and axial T1-weighted MRI scan demonstrating a syringobulbia and hydromyelia holocord and the enlargement of the ependymal channel greater than the 50% of the spinal cord's diameter. **D**, **E**, **F**, Postoperative follow-up MRI images showing a reduction in syrinx size.

tially limit long-term sequelae. Our case provides evidence that early lumbosacral lipoma removal before development of new subarachnoidal adhesions and maturation of fibro neural bands occur that complicate effective untethering could prevent additional surgeries. Additionally, at least in this case, the subsequent CSF obstruction leads to increasing the size of the distal syringomyelia that was resolved with shunting. In the literature, in the last 10 years, no cases of syringomyelia holocord associated with a lumbar sacral lipoma have been reported; hence, this paper offers a different surgical approach to this most difficult and hard to treat clinical problem.

■ Key Points

- The terminal "noncommunicating" syringomyelia in lumbar sacral lipoma has been reported to be associated with retethering in spinal lipoma in the 25% of the cases.
- The rise of distal syringomyelia isn't only linked to the kind of the spinal lipoma, but also to the difficulty to obtain the untethering and a smooth CSF flow between the subarachnoidal space and the ependymal canal.
- In patients with hydromyelia holocord superior than the 50% of the spinal cord's diameter is needed to perform a myelotomy to insert the catheter between the ependymal channel/syrinx and the subarachnoidal space to obtain the definitive resolution of the syrinx.

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