

Terminal Syringomyelia Mimicking an Intramedullary Tumor: Case Report

Intramedüller Tümörü Taklit Eden Terminal Syringomyeli: Olgu Sunumu

ABSTRACT

In this report, the authors present a 14-year-old male patient admitted with progressive paraparesis and urinary incontinence that developed in a 3-month period. Thoracolumbar MRI revealed an intramedullary lesion starting from the level of the T10 vertebra and extending to fill the sacral spinal canal. Based on the radiological findings an intramedullary tumor was suspected. Exploratory operation with right L5 hemilaminectomy was done and the intraoperative finding was syringomyelia which was punctured with releasing of the tethered cord. The patient was followed for 3 years with clinical and radiological regression of the pathology and no symptoms and signs have remained. Syringomyelia can mimic intramedullary tumors clinically and radiologically. That is why it is useful to consider exploratory hemilaminectomy to confirm diagnosis.

KEYWORDS: Hemilaminectomy, Intramedullary tumor, Terminal syringomyelia, Tethered cord syndrome

ÖZ

14 yaşında, 3 ay süresi içerisinde progresif paraparezi ve idrar inkontinansı gelişen bir erkek hastayı sunduk. Torakolomber MR'da T10 seviyesinden başlayan ve sakral spinal kanalı dolduran intramedüller lezyon saptandı. Radyolojik bulgular ışığında intramedüller tümör öntanısı konuldu. Hasta eksploratif sağ L5 hemilaminektomi ile opere edildi. Perop bulgusu olarak syringomyeli bulundu. Syringomyeli ponksiyon ile boşaltıldı ve gergin bant kesilerek kord serbestleştirildi. Operasyondan sonra hasta 3 yıl boyunca takip edildi. Semptomların tamamen kaybolması ile beraber tam radyolojik düzelme de görüldü. Syringomyeli klinik ve radyolojik olarak intramedüller tümörleri taklit edebilir. Bunun için hem tanıyı doğrulamak için, hem de tedavi amacıyla eksploratuar hemilaminektomi cerrahi yaklaşım olarak gözün önünde bulundurmak yararlıdır.

ANAHTAR SÖZCÜKLER: Hemilaminektomi, İntramedüller tümör, Terminal syringomyeli, Gergin omurilik sendromu

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INTRODUCTION

Syringomyelia is characterized by excessive accumulation of cerebrospinal fluid (CSF) within the spinal cord (13). Although concomitant hindbrain malformation exists with syringomyelia in the majority of cases (5), terminal syringomyelia in the lower third of the spinal cord is more commonly seen with spinal dysraphism, especially tethered cord (6,7). Syringomyelia also rarely develops in association with primary spinal intramedullary tumors (11,17) or intradural metastases (9). The pathogenesis, natural history and management of terminal syringomyelia remain controversial (18). Terminal syringomyelia can mimic intramedullary tumors; decision making for the approach and management can be more complicated in such cases. In this report we demonstrate the usefulness of exploratory hemilaminectomy to confirm diagnosis and even as a curative surgical approach for terminal syringomyelia, especially those associated with tethered cord.

CASE REPORT

A 14-year-old healthy boy presented with low back pain followed by progressive weakness of the lower extremities and urinary incontinence the developed over the past 3 months. On admission, the neurological examination revealed paraparesis which was more pronounced in the right leg (3/5), paraesthesia, paraparetic scissors-type gait with imbalance and sphincter dysfunction. There were no skin stigmata or spinal deformities. Thoracolumbar spinal magnetic resonance imaging (MRI) showed diffuse intramedullary lesion, with uncertain margins, which was hypointense on T1- and hyperintense on T2-weighted images with no contrast enhancement and extending from the level of the 10th thoracic vertebra to fill the sacral spinal canal (Figure 1A,B,C). Although no contrast enhancement was demonstrated, the expanding and the diffuse pattern of the lesion supported the radiological diagnosis of diffuse intramedullary tumor. Also, the diagnosis of fatty filum was radiologically excluded. Craniocervical MRI revealed no pathology. With the suspicion of the diagnosis of syringomyelia that extends through and expands the filum terminale, an exploratory hemilaminectomy was planned. Using a midline incision, the spinous process and the right lamina of L5 were exposed and a right hemilaminectomy was

performed. After exposure of the dura, a longitudinal dural incision was made and the sides of the dura were hanged. An expanded spinal cord obliterating the spinal canal was seen and punctured with an injection needle followed by evacuation of large amount of CSF. Drainage of CSF resulted in shrinkage of the spinal cord. The filum terminale was found tethered and upward withdrawal of the spinal cord was seen after releasing with microscissors. The operation was ended without any complications. Clinical improvement was observed in the early postoperative period. In the postoperative fifth day urinary incontinence disappeared and in the postoperative seventh day the patient started to walk without support with minimal weakness in the lower extremities. After 1 year, the patient's neurological examination revealed no neurological deficit and no urinary incontinence. Follow-up thoracolumbar spinal MRI obtained one year after operation demonstrated radiological regression of the pathology (Figure 2A,B,C). Three years after the surgery, the patient is still under the control of our outpatient clinic with no complaints and normal neurological examination.

DISCUSSION

Syringomyelia is caused by excessive accumulation of cerebrospinal fluid within the spinal cord (13). The pathophysiology of syringomyelia is still unclear although suggested theories may help us to understand the development and treatment strategies of this pathology. As suggested by Ball and Dayan (1) in 1972, there is probably an intracordal circulation of CSF from the subarachnoid space. It begins in the perivascular spaces (PVSs) and crosses the pia into the extracellular spaces in the cord and then the central canal. Rennels et al. (14) also thought that CSF in the central canal and spinal cord can move into the perivascular venous spaces and then to the subarachnoid space in a pseudo "lymphatic" system. From this point, movement of CSF through the arteriolar pial fenestrations, vascular space and interstitium to the central canal may occur (16). A return flow of fluid in the central canal or edema in the interstitial cord space to perivascular venous spaces in the cord may be produced by postural movement of the cord or straining, including the Valsalva maneuver. Syringomyelia was first classified by Barnett according to the relation between the cyst and the central canal into

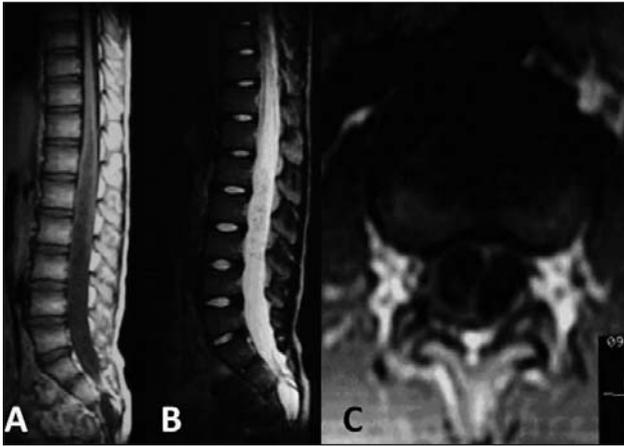


Figure 1: Preoperative lumbar spinal MRI demonstrating the diffuse intramedullary lesion, with uncertain margins, starting from the 10th dorsal vertebral level with extension and obliteration of the sacral spinal canal. A) sagittal T1-weighted, B) sagittal T2-weighted, and C) axial T1-weighted images.

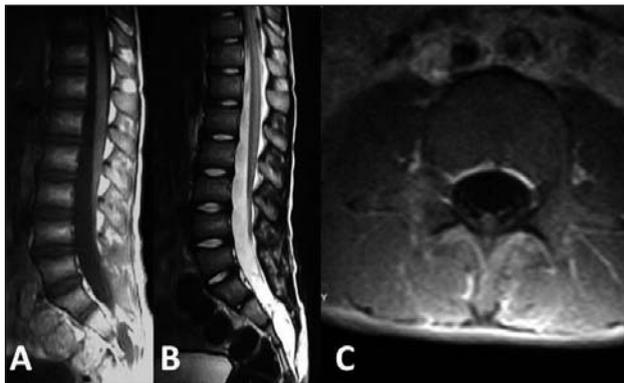


Figure 2: Lumbar spinal MRI obtained 1 year after the operation, showing no syringomyelia or tethering of the spinal cord. A) sagittal T1-weighted, B) sagittal T2-weighted, and C) axial T1-weighted images.

communicating (generally due to hind brain malformations) and non-communicating (due to spinal dysraphism) (7). In 1997, American Association of Neurosurgeons (AANS) had suggested a new classification for syringomyelia based on the etiology. In this classification syringomyelia is subgrouped into: 1) syringomyelia related to the fourth ventricle, 2) syringomyelia related to CSF circulation disturbances, 3) post-traumatic syringomyelia, 4) syringomyelia related to spinal dysraphism, 5) syringomyelia related to intramedullary tumor and 6) syringomyelia of unknown etiology. Although the major cause of syringomyelia is hindbrain malformation (5) (i.e. the communicating type), terminal syringomyelia in the lower third of the spinal cord is generally not

associated with hindbrain malformations and more commonly seen with spinal dysraphism (ie; non-communicating type), especially tethered cord (6,7). Terminal syringomyelia was present in 24% of cases of tethered cord (6), 27% of cases of occult spinal dysraphism (7), in 54% cases of meningocele manque and in 38% of split cord malformations (7). Many modalities are used for radiological diagnosis, such as intravenous and intrathecal contrast-enhanced CT scans and myelography, but MRI is still the gold standard of spinal neuroimaging. MRI, particularly contrast-enhanced, not only shows the exact location and extension of the intramedullary tumors but also allows the differentiation of tumor cysts and syringomyelia (8,17). Although MRI is the best radiological investigation for the diagnosis of syringomyelia, radiological findings are still not pathognomonic and syringomyelia can be confused with other pathologies. Surgical treatment of terminal syringomyelia is indicated in symptomatic cases. Although the optimum surgical procedure is still controversial, syringosubarachnoid shunt placement seems to be an effective treatment in symptomatic cases (3,4,10). In terminal syringomyelia associated with tethered cord, as in our case, spinal dysraphism or intramedullary tumor, the cyst may show regression or even disappear after surgical treatment of the underlying pathologies (2,10,12,15). Although there is a risk of recurrence in cases of the syringomyelia treated without shunting, this risk is known to be low after the definite treatment of the underlying pathology, and restoring the normal CSF flow (2,10,12,15). In the present case, the authors preferred only to untether the cord, thus achieving definite treatment of the underlying pathology and expecting a high rate of success in treating the syringomyelia (2,10,12,15), which may avoid the need for syringosubarachnoid shunting and the insertion of foreign materials. Also, in the present case the authors attempted to perform exploratory hemilaminectomy first, where the pathology was questionable. In cases of tumors, this approach provides a sufficient window for biopsy, thus; aiding us to plan the appropriate approach for further management, or can be enlarged to laminectomy or laminotomy if it is decided to remove the tumor in the same operative stage. On the other hand, in cases of syringomyelia it is a minimal invasive approach for draining the cyst and releasing the tethered cord without the need for

larger skin incision and bone exposure, thus; decreasing the risk of postoperative fibrosis and retethering.

REFERENCES

1. Ball M, Dayan AP: Pathogenesis of syringomyelia. *Lancet* 2: 799-801, 1972
2. Beaumont A, Muszynski CA, Kaufman BA: Clinical significance of terminal syringomyelia in association with pediatric tethered cord syndrome. *Pediatr Neurosurg* 43(3):216-221, 2007
3. Calderelli M, Di Rocco C, La Marca F: Treatment of hydromyelia in spinal bifida. *Surg Neurol* 50: 411-420, 1998
4. Chapman PH, Frim DM: Symptomatic syringomyelia following surgery to treat retethering of lipomeningomyeloceles. *J Neurosurg* 82: 752-775, 1995
5. Ellenbogen RG: Chiari 1 malformation. In Batjer HH, LoftusCM, eds. *Neurological Surgery: Principles and Practice Textbook of*, vol 1. Philadelphia: Lippincott Williams & Wilkins, 2003:960-967
6. Erkan K, Unal F, Kiris T: Terminal syringomyelia in association with the tethered cord syndrome. *Neurosurgery* 45: 1351-1360, 1990
7. Iskandar BJ, Oakes WJ, McLaughlin C, Osumi AK, Tien RD: Terminal syringohydromyelia and occult spinal dysraphism. *J Neurosurg* 83: 185-186, 1995
8. Kahan H, Sklar EM, Post MJ: MR characteristics of histopathologic subtypes of spinal ependymoma. *AJNR Am J Neuroradiol* 17:143-50, 1996
9. Keung YK, Cobos E, Whitehead R, Roberson GH: Secondary syringomyelia due to intramedullary spinal cord metastasis. Case report and review of literature. *Am J Clin Oncol* 20: 577-579, 1997
10. Koyanagi I, IwasakiY, Hida K, Abe H, Isu T, Akino M: Surgical treatment of syringomyelia associated with spinal dysraphism. *Child Nerv Syst* 13: 194-200, 1997
11. Lonser RR, Vortmeyer AO, Butman JA, Glasker S, Finn MA, Ammerman JM: Edema is a precursor to central nervous system peritumoral cyst formation. *Ann Neurol* 58:392-399, 2005
12. Ng WH, Seow WT: Tethered cord syndrome preceding syrinx formation – serial radiologic documentation. *Child Nerv Syst* 17: 494-496, 2001
13. Rekatte HL: Syringomyelia in children. In Batjer HH, LoftusCM, eds. *Neurological Surgery: Principles and Practice Textbook of*, vol 1. Philadelphia: Lippincott Williams & Wilkins, 2003:960-967
14. Rennels ML, Blaumanis OR, Grady PA: Rapid solute transport throughout the brain via paravascular fluid pathways. *Adv Neurol* 52:431-439, 1990
15. 19. Scatliff J.H, Hayward R, Armao D, Kwon L: Pre- and post-operative hydromyelia in spinal dysraphism. *Pediatr Radiol* 35: 282-289, 2005
16. Stoodley MA, Brown SA, Brown CJ: Arterial pulsation dependent perivascular cerebrospinal fluid flow into the central canal in the sheep spinal cord. *J Neurosurg* 86:686-693, 1997
17. Sun B, Wang C, Wang J, Liu A: MRI features of intramedullary spinal cord ependymomas. *J Neuroimaging* 13:346-51, 2003
18. Vandertop WP: Progression of terminal syrinx in occult spina bifida. *Child Nerv Syst* 19: 267, 2003