Syringo-subarachnoido-peritoneal Shunting for the Treatment of Syringohydromyelia

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ÖZET

Bu yazıda şiddetli suboksipital ve servikal ağrı yakınmaları olan ve sağ kolunda duyu bozukluğu bulunan, Chiari malformasyonu ile birlikte olmayan, bir siringohidromyeli olgusu sunulmuştur. Kistin sönmesi için hastaya siringo-subaraknoido-plevral şant uygulanmıştır. Ameliyat öncesi ve sonrası dönemdeki objektif ve subjektif klinik bulgular ile serviko-dorsal MRI bulguları tartışılmıştır. Hasta ameliyattan sonra hem nörolojik, hem de radyolojik olarak iyileşmiştir. Bu sonuç Chiari malformasyonu bulunmayan siringohidromyeli olgularındaki nörolojik bozuklukların siringo-subaraknoido-plevral şant uygulaması ile düzeltilebileceğini göstermektedir.

Anahtar kelimeler: Siringohidromyeli, sirinks, şant, tedavi

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SUMMARY

The authors reported a case of syringohydromyella without Chiari malformation whose symptoms were severe suboccipital and cervical pain and sensorial impairment in his right arm. Syringo-subarachnoido-peritoneal shunting was performed to collapse cystic cavity. Preoperative and postoperative clinical findings (both objective and subjective) and cervicodorsal MRI findings are discussed. The patient improved both neurologically and radiologically after surgery. This result indicated that syringo-subarachnoido-peritoneal shunting is effective in reversing neurological deterioration in patients with syringohydromyelia without Chiari malformations.

Key words: Syringohydromyelia, syrinx, shunt, treatment

INTRODUCTION

Cystic dilatation of the spinal cord was first described by the anatomist Estienne in the 16th century [4]. Olivier d'Angers coined the term syringomyelia and reported a case of syringomyelic patient whose spinal cord cavity was in continuity with the fourth ventricle [17].

In 1859, Stilling [17] showed that the central canal could persist into adult life and suggested the term hydromyelia. Barnett et al [3] classified syringomyelia according to its etiology:

1) Communicating syringomyelia (syringohydromyelia), which is associated with developmental anomalies at the foramen magnum and in the posterior fossa (e.g. Chiari type I and II malformations, Dandy Walker malformations, basiller anomalies);
2) Posttraumatic syringomyelia that develops as a late consequence of spinal cord injury;
3) Syringomyelia that results from spinal arachnoiditis;
4) Syringomyelia that is associated with spinal cord tumors; and
5) Idiopathic syringomyelia.

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CASE REPORT

This 30-year-old right-handed male patient presented with a 4-year history of severe suboccipital and cervical pain and sensorial impairment at his right arm. This pain was increased by coughing. Physical examination revealed decrease in temperature and pain sensation at the level of C3 C7 dermatomes and L'hermitte's sign was positive.

Non contrast enhanced thin sectioned CT scans of the cervical spine demonstrated hypodense cystic cavitation of the spinal cord at the level of C4-C6 vertebras. His cervicodorsal MRI investigation revealed syringohydromyelic cavities between level of C2-D11 vertebral spaces. The widest diameter of syringohydromyelic cavity was 1.5 cm at the level of D6 vertebrae (Figure 1).

Operation: Prone position was used for the exposure of the thoracic spine. After D6 total and D5 and D7 subtotal laminectomies were made, ligamentum flavum was removed and the dura was opened. High pressure cerebrospinal fluid streamed out when the cyst cavity was entered throuhg the posterior median sulcus. An infant size, distal slit valved, medium pressure shunt catheter was placed into the cavity. The shunt catheter's open and was inserted into the cyst cavity and multiple perforations were made on the proximal 2 cm length of the shunt catheter to connect the cyst cavity also to the subarachnoidal space. The shunt tube was secured to the dura and its distal slit valved and was put into the peritoneal cavity to complete the cyst-subarachnoid-peritoneal path.

Postoperative course: Immediate postoperative abdominal and thoracic X-ray films revealed correct placement of the catheter. His postoperative cervicodorsal MRI investigation showed collapsing of the cystic cavity after the successful syringo-subarachnoido-peritoneal shunting procedure (Figure 2).

DISSCUSSION

Defining the indications for operative intervention and deciding which surgical technique to perform are topics of considerably controversy. Numerous surgical interventions have been advocated for the

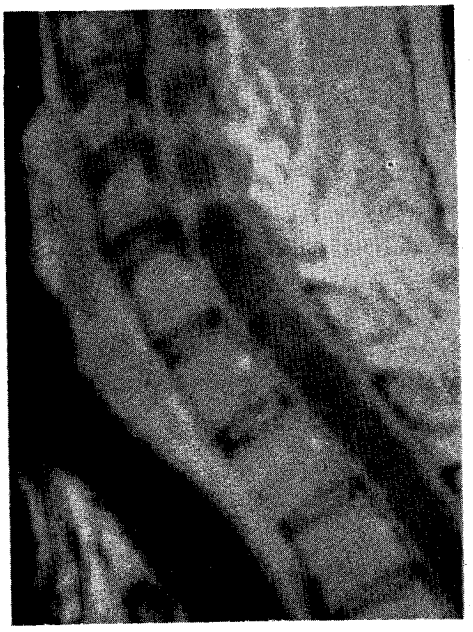

Figure 1. Preoperative MRI showing the syringo-hydromyelic cavity in the cervico-thoracic region.

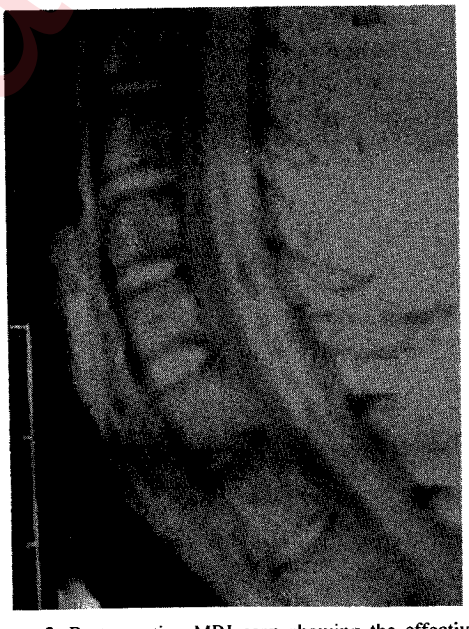

Figure 2. Postoperative MRI scan showing the effective shrinkage of the cavity and subsequent successful decompression of the spinal cord tissue.

treatment of hydromyelia since 1892 when Abbe and Coley [17] described improvement after laminectomy and aspiration of a syrinx. The various procedures can be grouped into four categories:

1) Decompression of the hindbrain malformation by suboccipatl craniectomy and upper cervical laminectomy [5,6,13,19,20];

2) Laminectomy and syringostom (myelotomy) [8,12,15,16,17];

3) Terminal ventriculostomy [19,21];

4) Percutaneous aspiration of the syrinx.

The improvement of shunt materials has revieved an interest in laminectomy, myelotomy and direct drainage of syrinx [15]. Love and Olafson [9], noted that the success of syringostomy depended upon maintaining a permanent fistula between the cyst cavity and the subaracnoidal space.

Syringo-subarachnoid shunting was advocated by Barnett et al [3], Shannon et al [14], and Tator et al [16]. Other authors suggested that shunting to a low pressure cavity such as the pleura or peritoneum is more efficacious [2,8,12,15].

Syringostomy along with shunting is the prefered modality for treating posttraumatic and idiopathic syringomyelia.

In our case we used syringo-subarachnoido-peritoneal shunting. We performed laminectomy at the region of maximal cystic dilatation and connected the cyst cavity both the subarachnoidal space and peritoneal cavity. In the early postoperative period, his suboccipital and cervical pain disappeared. The MRI investigation made in the early postoperative period showed the successful shrinkage of syringohydromylic cavity.

Anderson et al. [1] analyzed the time courses of 44 patients with syringomyelia: 7 (35 %) of 20 patients who did not undergo surgery had no progression of symptoms and were clinically stable for a median of 10 years after diagnosis. These investigators stated that although improvement was achieved after surgery, long term results were uncertaion.

Menezes [10] and Paul et al [11] reported excellent pain relief and improvement of motor function in 80-90 % of their patients with hydromyelia. Lesion et al. [8] reviewed 648 patients, of which 46 % improved, 32 % remained stable, 20 % had disease progression postoperatively.

CONCLUSION

This case report indicates that syringo-subarachnoido-peritoneal shunting reversed the signs and symptoms of neurological deterioration of our patient with syringohydromyelia in the early postoperative period. Longer follow up is needed before the success of this operation can be fully understood.

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