



Corpectomy - A Treatment of Last Resort in Post-Traumatic Ascending Syringomyelia in a Completely Paraplegic Patient

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Abstract

Post-traumatic progressive syringomyelia is a recognized complication of Spinal Cord Injury (SCI). In the cervical spine this can be life-threatening. Surgical options include decompression, adhesionolysis, shunting, myelotomy or rarely spinal cord transection or corpectomy. Treating the primary cause of CSF flow disruption is preferable and can lead to complete syrinx resolution. We describe performing a spinal cord transection in a 33-year-old patient with complete paraplegia. He developed a progressively symptomatic syrinx two years after traumatic SCI. Clinical and radiological resolution was achieved within one year of corpectomy. We advocate spinal cord transection or corpectomy in select cases with complete cord injury where tethering is disrupting CSF flow and causing ascending signs in particular bulbar symptoms.

Keywords: Spinal cord transection; Corpectomy; Post traumatic syringomyelia; Complete paraplegia; Ethical dilemma

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Introduction

Syringomyelia occurs in 0.5% to 4.5% of patients after traumatic cord injury; being twice as common in complete than incomplete cord injuries [1]. It can occur many years after cord injury; the syrinx cavity can progress superiorly or inferiorly, but more commonly ascends into normally functioning spinal cord [2]. The earliest symptom of syringomyelia is loss of pain and temperature sensation. This affects the arms and trunk, in a cape like distribution, caused by compression of decussating spinothalamic fibers. Enlargement of the syrinx can lead to anterior horn dysfunction, with an examination findings showing weakness, wasting and fasciculation. The dorsal columns are protected from the pressure of syrinx expansion so proprioception and light touch remain largely intact. Notably a late sign of syrinx cavity progression can be hyperhidrosis or Horner's syndrome [2], or bulbar dysfunction in the cervical-medullary region. Decompression of a syrinx can be achieved by shunting or myelotomy, with the added risks of damage to the normal cord, shunt failure and need for further surgery [3]. Alternatively, one can target the primary cause of syrinx formation with decompression (arachnolysis, untethering, and duraplasty). In extreme cases where neurological function has been completely lost below the tethered region, cord transection provides a useful addition to the neurosurgeon's armamentarium.

Case Presentation

Initial injury

We present a 33-year old patient injured in a motor vehicle crash in 2008. He suffered a traumatic brain injury and a Thoracic (T4) wedge compression fracture. After rehabilitation he returned to his baseline cognitive function. However, he remained completely paraplegic with a T2 sensory level and no sacral sparing; his injury was classified as having an American Spinal Injury Association (ASIA) score of "A". Upper limb function was preserved. The T4 fracture was treated non-surgically and achieved satisfactory fusion with moderate kyphosis at the level of fracture (Figure 1).

Neurological decline

After two years of stable neurological function, he developed progressive weakness in his upper limbs over a period of 7 months. Left arm power was reduced globally to 3/5 power on the



Figure 1: MRI T2WI Cervico-thoracic area demonstrates T4 Vertebral body fracture with spinal cord injury extending proximally and distally from this level: the cervical cord has normal cord signal.



Figure 2: MRI T2WI Cervical spine 2011 axial and sagittal sequences—significant syringomyelia involving entire cervical spinal cord and extending to the brainstem.

Medical Research Council (MRC) scale with no specific myotomal distribution. He lost the ability to independently roll his wheelchair and reduced hand dexterity made using buttons, zippers and cutlery very difficult. This was accompanied by dysesthesia of both arms, worse on left, without any specific dermatomal distribution. He deteriorated further, with swallowing problems and impaired cough reflex. Consequently, he had recurrent aspiration pneumonias requiring admission to the Intensive Care Unit (ICU) for ventilator support. Video-fluoroscopy was consistent with syringobulbar dysfunction. Further examination demonstrated altered speech (high nasal tone) and left deviation of the uvula. MRI at the point of profound deterioration identified an extensive multiloculated syrinx with involvement of the entire cervical spinal cord rostral to his previous injury with extension up to the medulla (Figure 2). This was not present on the MRI scan one year after the initial injury (2009) (Figure 1).

Operative management

The syrinx was a delayed complication of severe trauma to the spinal cord. Scarring at the level of the initial trauma resulted in obstruction of the normal CSF circulation at T4. This was complicated by multiple loculations within the syrinx (visible membranes inside the syrinx cavity) and spinal kyphosis at that level. Various treatment options were considered i.e. shunting of the syrinx, lysis of the adhesions to re-establish the circulation, and finally the option of corpectomy. This

has been performed successfully in paraplegic patients with complete cord injury [4-8]. Due to the progressive neurological deterioration, from a rostrally advancing multiloculated syrinx, in a patient with longstanding complete paraplegia without sacral sparing, the decision was made to perform a T3 spinal cord transection with duraplasty. A T3-4 laminectomy was performed. Midline dural opening revealed significant fibrosis and arachnoid adhesions around the spinal cord. Cord transection was performed at the level of fibrosis and scarring, just distal to the spinal cord with normal appearance. A section of the spinal cord measuring one centimeter was removed distally (cordectomy) to ensure discontinuity and prevent re-tethering from scar formation and fibrosis. Intraoperatively there was visible spontaneous drainage of the syrinx. An expansion duroplasty with watertight dural closure was performed, followed by posterior transpedicular spinal stabilization to prevent further deformity.

The transection of the spinal cord is demonstrated in the intraoperative (Video).

Post-operative outcome

The patient was repatriated to our spinal injury rehabilitation unit seven days post-op without complications. One-year post cordectomy, upper limb symptoms had returned to normal power bilaterally (MRC 5/5). He had a normal cough and a full functional resolution of his swallow. In addition, he no longer had any episodes of lower respiratory tract infection secondary to aspiration. The post-operative MRI scan at five months (Figure 3a) showed complete resolution of both the syrinx and brainstem edema. Sustained resolution was confirmed on MRI at five years' post op. (Figure 3b).

Discussion

Surgical decision making

Most literature emphasizes that the technique of cordectomy must be used only as a last resort and for a narrow list of indications such as end stage myelopathies, tethered cord syndrome, arachnoiditis and progressive spasticity [5]. In this case cordectomy was considered due to progressive, ascending neurological deterioration with an imminent threat to life. It must be offered only where there is total absence of function below neurological level and restoration of the subarachnoid space is not possible [7]. If there is still some function below the level of injury, then non-destructive procedures, such as decompression, syngo-peritoneal/pleural/subarachnoid shunts and myelotomy, should be offered instead.

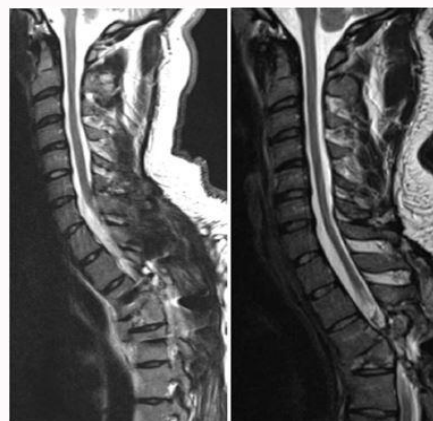


Figure 3: T2 MRI Sagittal sequences at (a) 5 months and (b) 5 years post cordectomy surgery with complete resolution of cervical spinal cord syringomyelia.

Surgical outcome

The main surgical goal of cordectomy is to prevent further neurological deterioration. Ewelt et al. [5] describe fifteen patients who underwent cordectomy: Eight had stabilization of their symptoms, six had improvement and one experienced deterioration. They were assessed using ASIA, Visual Analogue Score (VAS) for pain and a three-point scale for spasticity [5]. Cordectomy is highly effective at preventing progression of neurological decline in carefully selected patients [4,5]. Literature describes radiological resolution of syrinxes, and reduction in patient's pain and spasticity [8] and overall quality of life scores [9]. The more traditionally used shunting procedures were noted to have shortfalls and higher complication rates compared to cordectomy [6]. Literature therefore supports the use of cordectomy in carefully selected patients with clear indications as shown above.

Ethical Dilemma

Cordectomy is a procedure of last resort associated with potential psychological stress for the patient [9]. Due to the irreversible nature of the procedure patients should be psychologically supported and counseled prior to proceeding. At the time of writing, we had not come across literature on the psychological effect of cordectomy. It is also open to debate whether patients are permanently denied the opportunity for regeneration should such a breakthrough take place in the future. At the current moment, viable, functional regeneration of the central nervous system tissue has not been possible in humans. It remains a question of time whether this surgery will be deemed to have disadvantaged potentially curable patients. It is our recommendation to always discuss within a spinal MDT in presence of at least two qualified spinal surgeons, spinal injury/rehabilitation specialist, psychologist and whenever possible to involve the patient in the decision-making process.

Conclusion

Although ethically and psychologically challenging, cordectomy remains a useful surgical intervention to be used in cases of post-traumatic syringomyelia as a measure of last resort for patients with

progressive neurological decline. It forms part of the neurosurgeon's armamentarium for patients with complete paraplegia who develop progressive ascending syringomyelia. Appropriate patient selection and correct surgical technique are essential to prevent future surgical complications in this rare and complex patient group.

Video link: https://youtu.be/p3Xm_uGyBjE

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