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Clinical Case of the Month

Post-traumatic syringomyelia

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Post-traumatic syringomyelia is estimated to develop in more than 20% of individuals with traumatic spinal cord injury (SCI). The development can give rise to clinical symptoms 6 months to 26 years after the injury, and presentation 40 years post-injury has been seen by one of the authors.[1–4] We present an unusual case for comments and discussion.

Keywords: spinal cord injury; post-traumatic syringomyelia; tetraplegia; progressive symptoms

Case story

A forty-year-old man was involved in an accident as a passenger in a rally car, which rolled over several times. He was found to have fractures of the right 2nd and 3rd ribs with bilateral lung contusion, a fracture of the right scapula and right 2nd metacarpal bone. He was initially described as having normal sensation and movement of the legs. He was ventilated and sedated for the first 8 days. Due to urinary retention and generally decreased strength in the lower extremities additional investigations were carried out and a T3–4 fracture dislocation was diagnosed, with half a vertebral body slip anteriorly and laterally of T3 on T4. Thirteen days post-injury he was transferred to a neurosurgical department, where it was decided to continue conservative treatment of the fracture-dislocation. Sensation still was found to be normal, including in the sacral segments, there was normal voluntary anal contraction, but the muscle strength in the lower extremities was around 4, and co-ordination was impaired. A few days later the patient could empty the bladder normally. Five-and-a-half weeks post-injury he was admitted to the rehabilitation department, and discharged after a further 8 weeks of rehabilitation. At discharge he had normal voiding and defecation, and was walking nearly normally. He had some co-ordination problems and sensory disturbance of the left leg, and a 10 cm broad band with dysaesthesia on the left thorax. He was not followed up after discharge.

Approximately 8 years later he noticed light sensory disturbances first in the right and later the left C_8 segment. An MRI was performed repeatedly over the next 3 years and showed, apart from the known fracture, a lobulated syrinx from the level of the fracture to the upper border of C5. In addition the MRI showed a 3 cm long intramedullary cyst at the level of T5. Fourteen years post-injury his symptoms had progressed with disturbed sensibility in all fingers except the thumbs, increasing co-ordination problems in the right leg, and urinary retention necessitating intermittent catheterisation, together with loss of ejaculation, impaired erection, and increasing difficulty in controlling defaecation. An MRI showed the syrinx had increased in size cranially, with only minimal medullary tissue remaining (Figure 1). Fifteen years post-injury he was transferred to our department and a syringo-peritoneal shunt was performed. At the time of operation he was almost anaesthetic and analgesic below T_4, but he could walk without aids. After the operation there was some regression of the sensory disturbances in the hands. Three months after operation he experienced increasing spasticity and difficulty in walking. MRI showed the syrinx had collapsed after the operation, and no further neurosurgical intervention was indicated. In the following months decreasing strength in the lower extremities made even standing impossible, later finger flexion and abduction decreased bilaterally (strength 3–4) making ADL difficult, and sensation

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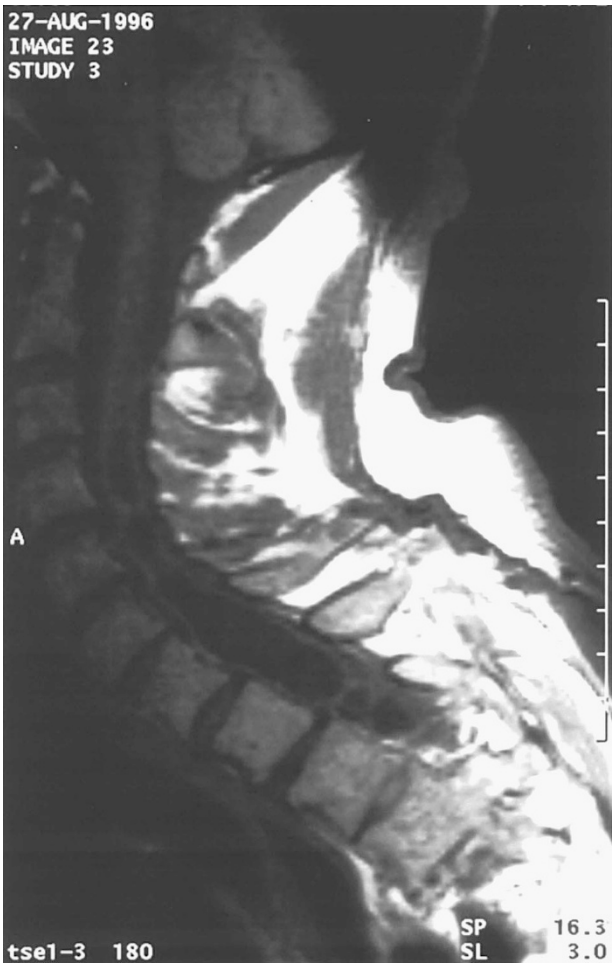

Figure 1 MRI of the cervical and upper thoracic spine. There is a fracture of T4 with an anterior dislocation of T3. A lobulated syrinx is seen from C3 to T3

was abolished from T_2. A further MRI nearly 16 years post-injury was unchanged compared to the previous one half a year earlier. Over the following months the neurological situation stabilised, and he presented with a C_{7-8} complete tetraplegia with severe lower limb spasticity, and a denervated urinary bladder.

Comment no. 1

U Bötel

Since MRI enables us to study medullary changes in the course of time following severe vertebral trauma, post-traumatic syringomyelia is discovered not so rarely, although the patho-mechanical factors are not fully understood. Decreased or absent CSF circulation over the traumatised segment of spinal cord may play an important and decisive role either by post-traumatic arachnoiditis or by stenosis and kyphotic deformity of the spinal canal.

In this case, apparently an unstable fracture of a flexion-rotation type of T3 and T4 led to a severe kyphotic deformity of the upper thoracic spine resulting in a marked cervical hyperlordosis, which was responsible for the extended syringomyelia of the cervical cord and the late neurological deterioration at least in the C_{7-8} segments. The complete spastic paralysis of trunk and lower limbs was probably due to a late post-traumatic myelopathy in connection with the kyphotic deformity and mechanical irritation of the injured cord, as signs of an incomplete and improving paraplegia had already been present immediately after the trauma. The latter proposal is supported by the fact that the main neurological deterioration was in the long tracts, whereas the short tracts are more influenced by the pressure of the syrinx.

I would have recommended the restoration of the thoracic spinal canal and reconstruction of the normal alignment including the restoration of CSF passage by duraplasty and additionally shunting the syringomyelia at the earliest possible time. Ideally the operative reconstruction of the thoracic spine should have been done after the phase of intensive care treatment, or at least at the first signs of neurological deterioration. Meanwhile a reconstruction will be of benefit.

Comment no. 2

BP Gardner

This fascinating and disturbing case study indicates how little we understand of the patho-physiology of post-traumatic spinal cord syrinx.

Although it is tempting to speculate that earlier neurosurgical intervention when the syrinx first became symptomatic would have prevented the progressive deterioration, there is no certainty that this would have been the case. Although neurosurgical intervention in such cases usually prevents further deterioration and very occasionally gives rise to neurological improvement, sometimes there is further neurological loss that does not always recover. SCI persons with mildly symptomatic syrinxes faced with this uncertainty frequently opt for a conservative strategy. Very often this is successful. There are many SCI syrinx patients with mild clinical features who either remain neurologically static or improve spontaneously. Conversely the reverse may occur with neurosurgical intervention. Earlier this year for example an ambulant patient under my care became almost completely paraplegic following a syringopleural shunt. Fortunately he has since recovered almost to his pre-operative state. In our study[4] we demonstrated that the incidence of syrinx formation in thoracic level SCI persons patients rose from 30% at 20–29 years to almost 50% at 30–39 years post-injury. The majority were asymptomatic. The longer the syrinx the greater was the chance that it was symptomatic, though some very large syrinxes were asymptomatic. There was no relationship between the

prevalence of syrinx and the degree of spinal canal compromise or angulation adjacent to the level of injury.

In the absence of convincing data on outcome my approach to SCI patients with asymptomatic syrinxes is to advise the avoidance of situations that give rise to abrupt physical stress, as these are associated with clinical deterioration, and to monitor the patients regularly and carefully, both clinically and by MRI scanning. Those who develop progressive neurological symptoms and signs are advised to undergo surgery. On this basis I would have offered this patient surgery earlier than 15 years post-injury. If he had declined I would not have pressed him as some patients spontaneously cease to deteriorate whilst some others deteriorate with surgery.

A very carefully and well-conducted prospective longitudinal multi-centre study is requried to identify the natural history, both clinical and radiological, of this ill-understood condition. Perhaps in this way the optimum method of treatment in any individual will become more apparent.

Comment no. 3

J Little

In this case of incomplete T_4 paraparesis, ascending sensory loss in the hand was noted 8 years after spinal cord injury, attributable to an ascending syrinx. Motor impairments appeared at 14 years as right leg incoordination and at 15 years as hand weakness, the syrinx now ascending to the medulla. Weakness commonly lags behind syrinx elongation as revealed by MRI and sensory loss. This delayed onset of weakness leads to delayed awareness by patient and physician that the post traumatic syrinx is causing continuing, subclinical neurologic damage.

The lag in onset of weakness may be due to several factors. First, syrinx location at the base of the dorsal horn directly damages sensory relay neurones of the spinothalamic tract; only marked syrinx dilation damages anterior horn motorneurones and descending motor axons to lower limbs at the white matter periphery. Second, gradual alpha motorneurone loss and descending motor axon loss are supplanted by continuing axon sprouting of spared motor-units and spared cord descending motor axons. In addition, the weakness develops over many years; both patient and physician sense decline only later. Finally, our standard clinical measure of motor function, the manual muscle test, is insensitive to strength changes in the above antigravity strength range.

Because of delayed overt weakness, patient and clinician may opt not to intervene. By the time weakness is apparent, it may be irreversible. A useful clinical strategy may be to serially quantitate strength, measure motor-unit amplitudes with quantitative electromyography and measure motor evoked potentials to attempt to identify ongoing subclinical neurologic decline. This may allow intervention before irreversible weakness develops.

However, treating syrinxes is problematic, particularly in those with spared descending motor axons. These patients with incomplete spinal cord injury commonly lose the ability of walking, either immediately or soon after shunting. The reason is unclear. Perhaps shunts impose harmful mechanical forces on descending motor axons. Early shunting only precipitates early loss of walking, in an attempt to save upper limb function. This trade-off is usually not acceptable to patients.

New treatments are needed that provide long-term decompression of the syrinx and that protect existing function above and below the syrinx. Subarachnoid space reconstruction or neural tissue grafts are two possible alternatives but their long-term success rates are not known. I hold out hope that through better understanding of the pathophysiologic mechanisms causing syrinx dilation, we will be able to devise effective non-operative treatments.

Comment no. 4

H Ohta

It is widely accepted that the realistic goal of the surgical treatment for post-traumatic syringomyelia is improvement of symptoms and stabilisation of neurological status. From my experience of 33 cases with post-traumatic syringomyelia, it was observed that the shunting was effective to improve neurological symptoms when it was done within a year after neurological symptoms had started to progress. However, when the operation was done more than one year after progression of symptoms, the operative effect was only limited to stabilise the neurological status.

I have not had such a severe case of deterioration as here demonstrated, but I have experienced two mild cases of delayed deterioration, even though the syrinxes had collapsed. I suppose that these were too late to be operated on (one was 8 years and another was 29 years after progression) and furthermore they had much more extensive arachnoiditis than other improved patients.

Therefore, I consider that patients with post-traumatic syringomyelia should be candidates for surgery as soon as neurological symptoms start to progress. In the presented case shunting of the syrinx was performed 7 years after the neurological symptoms started to deteriorate. I am afraid, it was too late to operate.

Another thing is, that I am wondering why a syringo-peritoneal shunt was selected in this case. Because I understand, that a syringo-peritoneal shunt tube is larger and more invasive than a syringo-subarachnoid shunt tube. Regarding the operative procedure I recommend performing a one or two level laminectomy, then inserting a pair of syringo-subarachnoid shunt tubes into the subarachnoid space, one

up and the other further down from the injured spinal cord.

It is suggested, that the causes of delayed deterioration in this case might be arachnoiditis and subsequent spinal cord ischemia or obstruction of the shunt tube. The shunt operation could accelerate the pre-existing arachnoiditis. It has been reported that the spinal cord at the level of T_{2-3} is most vulnerable to ischemia, because of the watershed of the blood supply to the spinal cord.

Comment no. 5

RD Shrosbree and R Melwill

The MRI image has come through rather badly, so comments are not made with a clear knowledge of the neuro-radiological evaluation of the patient.

Conservative management of the T3-4 fracture dislocation suggests that there may have been residual disruption in the spine alignment with possibly a kyphosis at the site. This together with residual arachnoiditis at the fracture site probably accounts for the development of the syringomyelia. Sgouros and Williams[3] stress the importance of a kyphotic deformity in the evolution of syringomyelia, as does Batzdorf *et al*[5] in his recent critical appraisal of shunting procedures for syringomyelia.

Placing shunts in the syrinx has been in vogue for long enough for some interesting long term studies to emerge. The operation does not necessarily provide the answer that a simplistic explanation of the problem would have presupposed. Batzdorf *et al*,[5] reporting on a series of patients from the University of California at Los Angeles Medical Centre, has shown shunt failure in 46% of cases. In the case under discussion the shunt system functioned adequately, yet there was a continued deterioration in the patient's neurological state. Other factors must play a part in the patient's condition. Is this not associated with tethering of the spinal cord at the site of the injury or alternatively at the site of the shunt operation? This has been mentioned in the past by Sgourus and Williams[3] and by Steinmetz.[6]

This case reaffirms the contention that cyst drainage alone is not the solution to the treatment of syringomyelia. Must we look to the correction of post-traumatic kyphosis early, and to ensuring that there is no tethering of the spinal cord by arachnoid adhesions and other structures in both the treatment and prevention of syringomyelia?

The only point we can add is that it is indeed unfortunate that he was not referred earlier. In our experience the sooner one treats proven cases of syringomyelia and cysts with functional effects the better the results. With a 'retrospectroscope' and the subsequent history this may well have been the course to follow. Although my experience with this condition is relatively limited, the few cases that we have had operated on early have done well and often regained what they had lost.

Recently however a colleague who is paraplegic had a repeat operation after a successful initial operation 3 years previously. This however did not stop progression of symptoms and a further operation was done to drain the syrinx further down and release a tethered cord. Unfortunately his condition did not improve and he is now left as a C_7 tetraplegic.

In summary therefore, perhaps the best approach would be early surgery as soon as there is functional loss, in addition to untethering of the cord.

Discussion

The patient presented developed sensory disturbances 8 years post-injury and a MRI showed a lobulated syrinx cranially to the fracture-luxation at the level of T3-4. Six years went by without significant progression, but 14 years post-injury the patient experienced progression with further loss of sensation, co-ordination problems and sphincter symptoms. MRI showed the syrinx had increased in size cranially. Fifteen years post-injury a syringo-peritoneal shunt was inserted. Three months later the patient lost almost all motor function in the legs and sensation was abolished from T_4. MRI showed a collapsed syrinx.

Except for Gardner the authors consider it advisable in the acute treatment of a case presenting like this to perform surgical reconstruction of the normal alignment and hereby restoration of CSF passage.

Perrouin-Verbe *et al*[7] in their study found a significant correlation in the occurrence of post-traumatic syrinx-formation with spinal canal stenosis in both the sagittal and axial plane. This correlation was not found in the series of Wang *et al*.[4]

All commentators opt for earlier surgical intervention on the syrinx than was done in the case, ie when the first sensory disturbances appeared 8 years post-injury. At that time the patient did not have any motor-symptoms. It may be too late to save motor function if one waits for motor symptoms to appear before shunting the syrinx.

Syrinxes seem to cause more harm to the spinothalamic tracts than the descending motor axons and patients' first syrinx symptoms are therefore often sensory disturbances and only later (years later in this case) the development of motor-symptoms.

Bötel remarks that neurological deterioration might be derived from multiple reasons not only relying on the intramedullary cyst.

Little and Gardner describe cases, like the one presented, with successful shunting where MRI showed a collapsed syrinx but with disappointing progression of motor deficits. The reason may be mechanical pressure from the shunting tube, altered vascularization of the medulla, sudden changes in the pressure, scar formation, spinal cord tethering or a combination of these.

Shunting procedures have been and probably still are the most widely used treatment of post-traumatic

syrinx formation. Shunt failure due to glial obstruction, migration and disconnection is seen more often in the treatment of syringomyelia than after shunting hydrocephalus. Batzdorf *et al*[5] report a 59% incidence of shunt failure where the patients developed recurrent cyst expansion. Shunt obstruction was the reason for most of the failures.

Ventureyra *et al*[8] reported the innovative use of a myringostomy tube for syringostomy. This very small device is thought to reduce scarring compared with ventricular catheters. Long-term follow-up will show the effectiveness of this technique.

Sgouros and Williams[3] advocate decompressive laminectomy and formation of a surgical meningocele and state that drainage procedures have little place in the management of syringomyelia.

Transplantation of foetal neural tissue into the syrinx is another technique,[9] but long-term observations are not yet available.

Long-term monitoring of patients with spinal cord injury is essential if treatment is to be optimal. Preferably the patients should be followed longitudinally by experts who are able to monitor clinical signs which could be indicative of formation or progression of a syrinx. Developing clinical symptoms like pain, altered sensibility, impaired motor function, increased spasticity and sweating are well known indications of possible formation or expansion of a syrinx.

With the presented experience we suggest that in the future all spinal cord injury patients should have an MRI scan 6 months after the trauma, independent of their recovery from the injury. If they have any syrinx-formation they should be followed closely for clinical indications of progressive syringomyelia irrespective of their functional level, ie even persons considered to have complete recovery should be controlled regularly in the spinal cord centre. Because syrinx formation can commence at any stage following spinal trauma regular subsequent MRI scanning is required.

Post-traumatic syringomyelia is indeed a difficult issue, both when discussing its pathophysiology and its treatment. In spite of the wide range of treatment possibilities, none of them can be characterised as more than 'fair'.

The solution to a better treatment of post-traumatic syringomyelia in the future undoubtedly rests in a better understanding of the pathophysiology. As stated by Gardner, a very careful and well-conducted prospective longitudinal multi-centre study is required to identify the natural history, both clinical and radiological.

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