Review article

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Posttraumatic syringomyelia

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Abstract: Posttraumatic syringomyelia is an uncommon cause of late neurological deterioration in a patient with spinal cord injury. The pathology has received recent attention due to increased physician awareness and the availability of MR imaging. The underlying mechanism is probably local arachnoid scarring and obstruction to CSF flow, followed by seepage of CSF through the Virchow-Robin spaces. Persistent deformity predisposes to formation of posttraumatic syrinx. Anatomical decompression, division of arachnoid scarring, correction of spinal deformity and shunting procedures are measures to arrest the progress of the syrinx.

Keywords: spinal cord, spinal injury, syringomyelia.

INTRODUCTION

Post-traumatic syringomyelia is a clinical syndrome of delayed progressive myelopathy often corresponding to spinal segments distant from the level of original injury. Harvey Cushing described a syringomyelic syndrome in a patient with cervical gunshot wound[1]. He viewed hematomyelia as the pathologic substrate of both – acute and delayed post-traumatic syringomyelia. The exact incidence is unknown, since the manifestations occur in few patients following spinal cord trauma, and there is a paucity of well-studied autopsy cases of post-traumatic spinal cord cavitation. Wagner & Stolper in 1898 provided the first autopsy description in the post-mortem study of a patient with fractures of fifth thoracic and first lumbar vertebrae[2]. They described extensive cavitation of the cord, both rostral and caudal to an area of contusion, which was in close proximity to an area of leptomeningeal thickening, lined by glial cells, and filled with turbid gray fluid. Holmes described cylindrical cavities adjoining the injured segment, extending four or five segments cephalad or caudad[3]. The incidence of spinal cord injury in the developed countries varies between 11.5 and 53.4 cases per million population[4]. In the United States, the incidence is between 721 to 906 per million population. Nearly 21% to 28% of these patients will have syrinx if investigated between one to 30 years after injury[5]. Symptomatic syringomyelia is however seen in only 1% to 9% of the spinal injury population, and in conformity with the gender distribution of the injury, is seen more often in young males[5,6]. Higher association has been observed in patients with complete lesion[7]. In a prospective study involving 449 spinal cord injury patients followed yearly for a 6-year period found that 4.45% of the patients developed a symptomatic syrinx[8]. Over the past few decades, there has been improvement in the acute management of spinal cord injury, improved rehabilitation and long-term survival. This improvement coupled with availability of imaging modalities, has made post-traumatic syringomyelia a more prevalent and well-recognized cause of delayed neurological deterioration in patients with spinal cord injury.

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THE TERMINOLOGY

Posttraumatic syringomyelia is an important cause of delayed neurological deterioration that extends rostral to the level of the initial injury. There were early reports of "syringomyelic syndrome" preceded by trauma, there was no confirmation of the pathological lesion[9]. Charcot in 1892, Lloyd in 1894 and Cushing in 1898 described syringomyelic syndrome soon after the injury to spinal cord[1,10,11]. That trauma was causally linked to the formation of syrinx and late deterioration of residual neurologic function following spinal cord trauma was eventually established after improvement in myelographic techniques. Late deterioration of neurologic function has since been termed variously as, besides posttraumatic syringomyelia, ascending cystic degeneration of the cord, syndrome of chronic injury to the central cervical cord, posttraumatic progressive myelopathy, posttraumatic cystic myelopathy, and progressive posttraumatic cystic myelopathy, and porogressive posttraumatic myelomalacic myelopathy (PPMM)[12] (Falcone et al). In

PPMM however, there is no cavitary lesion in the cord, but a zone of myelomalacia with widening and posterior or lateral tethering of the spinal cord. MRI shows irregular, ill-defined margins of myelomalacia that do not follow CSF intensity. Thus, posttraumatic syringomyelia has to be distinguished from post-traumatic cavities due to myelomalacia that extend over less than two cord segments, and are non-progressive in nature. The timely diagnosis and appropriate surgical measures is critical to arrest the neurological worsening and to preserve function.

PATHOGENESIS

The pathophysiologic basis for the formation and extension of syrinx in the injured cord remains uncertain. The information available so far has been based on clinical observations and retrospective studies. The lack of scientific method to test a hypothesis derived from clinical observations about post-traumatic cavitory lesions in the spinal cord might imply that the inference drawn from various studies may be untested conclusions. Relevant animal models would overcome this deficiency. Once the suggestion was that all syringomyelias – occurred due to an embryonic substrate[13,14] in the form of a small posterior fossa. This meant that the patients with spinal cord injury who developed syringomyelia developmentally had small posterior fossa. A communication from the syrinx with the fourth ventricle had been demonstrated in reports by Oakley et al and McLean et al[14,15]. In autopsy study conducted by Milhorat et al, the syrinx lining after trauma had different hisopathologic picture (including chromatolysis, Wallerian degeneration, focal necrosis in the cells and nuclei of the cord) as compared to that from the syrinx associated with Chiari malformations[16]. Williams et al rejected the congenital substrate concept after Nurick et al demonstrated absence of any anomalies of the posterior fossa in two of their patients[17,18].

Posttraumatic syringomyelia is regarded as a two-step process: formation of a cyst, and is extension by secondary factors. The intial cystic lesion results from multiple factors that occur after trauma. These are mechanical damage, local ischemia[19] due to arterial and venous obstruction, liquifaction of hematoma, lysosomal and other intracellular enzymes[20,21]. Experimentally, it has been shown that spinal cord cavitation occurs after intraspinal injection of quiscolic acid, an excitatory amino acid (EAA) agonist, which results in cell death[22]. This excitotoxic EAA activity could be implicated in the formation of cavity after injury. Mechanical damage followed by subarachnoid scarring plays an important role in the initiation of the cystic lesion, which subsequently extends cranially and caudally[23,24]. Partial or complete obstruction to CSF flow at the injury site leads to further extension of the syrinx. Initially, extension was thought to be due to a valvelike connection of the cavity to the subarachnoid space[25]. There is possibly a unidirectional flow of CSF from the spinal subarachnoid space into the posttraumatic cyst through the Virchow-Robin spaces[26], and this flow is an important factor in the extension of the syringomyelic cavity.

Severity of the cord injury too is probably unrelated to the formation of syrinx[27,28]. Trivial injury to the cord was as likely to result in syrinx as a severe injury. In a review of 21 patients with syringomyelia out of 75 patients with spinal cord injury, Perrouin-Verbe et al evaluated the relationship between stenosis of the spinal canal caused by insufficiently reduced spinal fractures and occurrence of posttraumatic syringomyelia[29]. They found higher incidence of syringomyelia in patients with stenosis. Similar results were reported by Abel et al when they evaluated 207 spinal-injured patients[30].

PRESENTATION

Symptoms may manifest soon after injury[1] or may be as late as 33 years[31]. Approach involves a detailed history and clinical examination. Pain is a frequent symptom, the incidence ranges between 63% and 89%[7,9]. Increased motor deficit was seen in 63% (Rossier, Vernon), while dissociated sensory loss was seen in 87% [6]. Other features may be hyperhiderosis, autonomic dysreflexia, Horner's syndrome and respiratory insufficiency. Signs and symptoms may be unilateral or bilateral, and may alternate from side to side with changes in position. Syrinx may extend into the medulla, and involvement of cranial nerves should be recorded. Multimodality neurophysiologic assessment can complement clinical examination. This includes electromyography, SSEPs and motor evoked potentials for evaluation of the afferent and efferent spinal cord pathways plus the local H-reflex.

IMAGING

MRI is the diagnostic study in a patient suspected of having posttraumatic syringomyelia. Studies should include T1- and T2-weighted images. Intravenous gadolinium should be given to distinguish scar tissue. MRI-compatible implants in stabilizing the acutely injured spine have followed use of MRI in the evaluation

of spinal cord injury. However, many of the patients operated upon in the era before MRI became uniformly available, will have MRI-incompatible implants. These patients will require high-resolution computed tomography with intrathecal contrast. In these cases, delayed scans are obtained 6 to 24 hours after the injection. Real-time ultrasound, especially in thin individuals, is useful in post-laminectomy follow-up studies.

MANAGEMENT

Management of posttraumatic syringomyelia encompasses two aspects, namely, drainage of the syrinx, and correction of deformity. Surgical planning requires assessment of the site of CSF obstruction, the need for decompression, untethering of the cord in the subarachnoid space, deroofing of any subarachnoid cysts, and the availability of the spinal subarachnoid space for drainage of the syrinx. The approach may have to be posterior, for decompression and lysis of arachnoid adhesions, and placement of syringo-subarachnoid shunt, followed by anterior approach for correction of deformity[32].

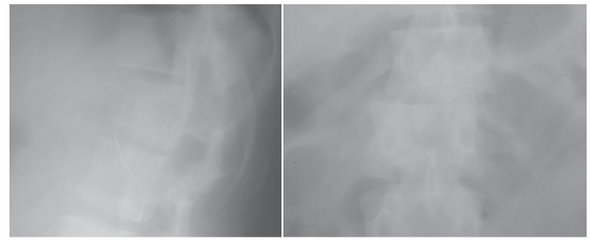
ILLUSTRATIVE CASES

CASE 1

A 27-year-old army soldier was admitted in 2000, with persistent backache following a fall from a height. He was continent, and had no neurological deficit. Spine radiographs revealed wedge compression fracture LV1 (Figs 1a & b). He was managed by decompressive laminectomy alone followed by ambulation with a brace, with relief in backache. He was readmitted in 2008, with increasing dysesthesiae over the left side of trunk and stiffness of left lower limb. Tendon jerks were brisk and he had dissociated sensory loss over the trunk on the left. He had difficulty in voiding completely. Spine radiograph showed worsening of deformity (Fig 2). MRI showed a holocord syrinx extending to the medulla (Figs 3 & 4). He underwent spinal cord decompression and lysis of arachnoid, followed by syringosubarachnoid shunt. There was relief in dysesthesiae and improvement in continence. MRI done after three months showed reduction in the size of syrinx.

CASE 2

A 19-year-old female patient sustained severe dorsolumbar spine injury in a high-speed motor vehicle accident and



Figs 1a & b: Dorsolumbar Spine radiograph (lateral) showing compression fracture LV1

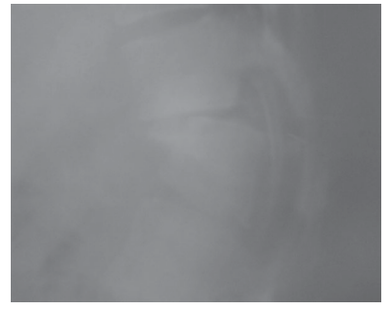


Fig 2: Dorsolumbar spine radiograph after eight years showing worsening of deformity

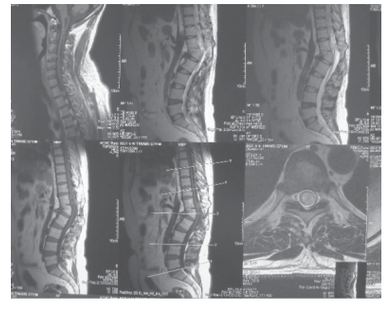


Fig 3: MRI (T1-weighted sagittal image) showing stenosis at LV1 and holocord syrinx

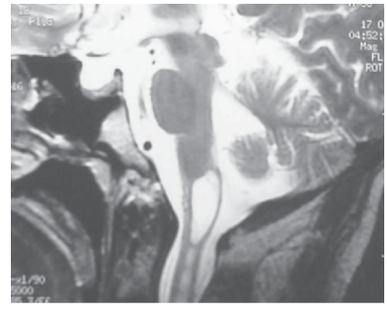


Fig 4: MRI (T2-weighted sagittal image) showing bulbar exension of the syrinx

was admitted with paraplegia (Frankel A). CT and MRI of dorsolumbar spine revealed complete dorsolumbar disjunction, and cord transection (Figs 6 & 7). Cervical

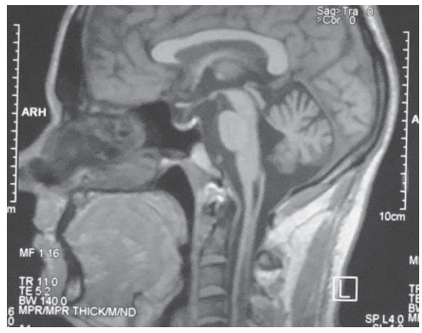


Fig 5: MRI (T1-weighted sagittal image) showing reduction in the syrinx after syringosubarachnoid shunt

Fig 6: MRI showing fracture dislocation at D12-LV1

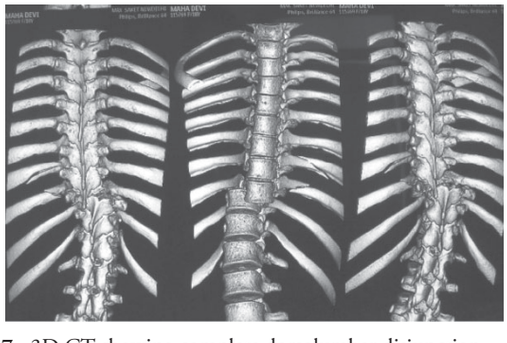


Fig 7: 3D CT showing complete dorsolumbar disjunction

cord was normal (Fig 8). She was managed with steroids, back care, intravenous fluids and analgesics. Twenty-four hours later, she developed respiratory failure, requiring ventilatory support. MRI cervical spine revealed syrinx extending the entire extent of the cord (Figs 9, 10a & b). She died 18 hours later due to respiratory failure.

Fig 8: Cervical MRI showing no evidence of syringomyelia

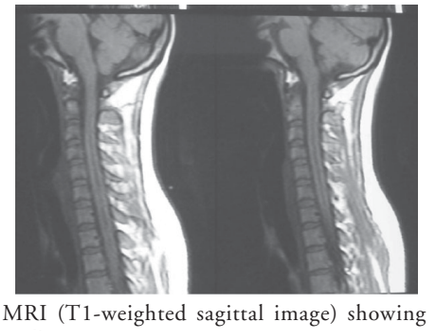


Fig 9: MRI (T1-weighted sagittal image) showing cervical syringomyelia

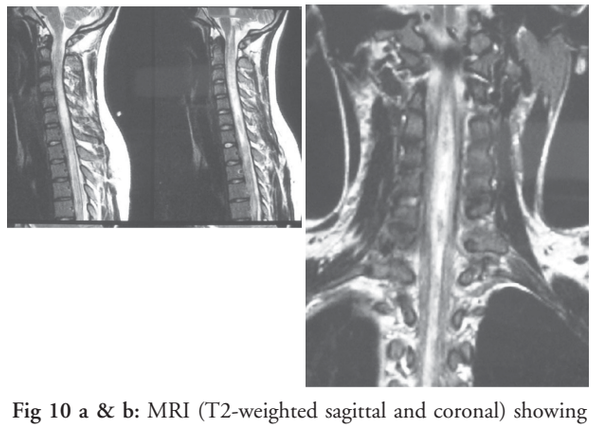


Fig 10 a & b: MRI (T2-weighted sagittal and coronal) showing cervical syringomyelia

CONCLUSION

Posttraumatic syringomyelia is increasingly being recognized as an important cause of delayed neurological deterioration after spinal cord injury, mainly due to availability of MRI for diagnosis and follow-up. Intradural obstruction to CSF flow and external spinal deformity

along with an injured cord are the anatomical substrates for the formation of syrinx. Management involves intradural decompression, shunting the syrinx and correction of deformity. There is relief in sensory symptoms and arrest of motor worsening.

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