

Case Reports

Syringomyelia – As a Late Complication of Tuberculous Meningitis

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Summary

The aim of this paper is to demonstrate the unusual MR features of thoracic syringomyelia following TB meningitis and to discuss the neurosurgical aspect of the treatment of this rare entity. Four years after a TB meningitis episode, a 30 year-old female patient developed a progressive spastic paraparesis. MR studies revealed multiloculated syrinxes throughout the thoracic cord. She had a syringo-subarachnoid shunt with a silastic “T” tube inserted. On the first postoperative day, she showed a dramatic neurological improvement, but unfortunately her paraparesis progressed to the preoperative level within a month despite diminished size of the syrinxes on the control MRI examination. Two and a half years after the operation the patient complained of having a burning type of central pain, and further deterioration in neurological function. Thoracic spinal MRI examination demonstrated enlarged syringomyelic cavities. At the second operation syringo-peritoneal shunt insertion was performed via right T_{10–11} hemilaminectomy using a “T” tube. At present, 4 months after the second operation, the patient’s neurological examination demonstrated decreased spasticity, and improved strength in the legs compared to the preoperative level. MRI is the first choice of investigation in detecting TB related myelopathy as it provides a greater detail of pathological changes within and around the spinal cord such as syrinx formation and arachnoiditis. The MR findings are also helpful in deciding the management and predicting the outcome. Presence of multifocal loculations and arachnoid adhesions is the likely cause of treatment failures and poor prognosis.

Keywords: Syringomyelia; tuberculous meningitis.

Introduction

Tuberculosis remains a major health problem worldwide. Non-hindbrain related syrinx as a complication of meningitis is well documented. The present paper aims to present a case who developed post meningitic arachnoiditis resulting in the generation of syringomyelia. Syringomyelia secondary to TB meningitis is an exceedingly rare condition of which the relevant data revealed only 23 cases of this type [1, 3, 4,

6, 8, 9, 11, 13, 14, 15, 17, 18, 19, 20]. The radiographic diagnosis is best made by MR. As these cases are usually associated with extensive arachnoid adhesions and multifocal loculations, surgical attempts generally fail to improve the neurological status. If there is any treatable cause of neurological deterioration such as syringomyelia every attempt should be made. In post-meningitic syringomyelia, syringo-peritoneal shunt insertion may give satisfactory results despite the duration of the neurological deficits.

Case Report

A 30 year-old female patient was admitted to our clinic due to the complaint of progressive weakness in both legs. She was a former patient of our hospital, who was hospitalised six years ago (April 1992), with a diagnosis of tuberculous meningitis. She was then treated with the four antituberculous drug regimen (rifampicin, isoniazid, pyrazinamide and ethambutol) for 18 months and was rehabilitated. Over the subsequent two years, she had a substantial recovery except for the complaint of minimal weakness in both legs, which prompted her physician to investigate the spinal cord. The relevant MRI revealed the presence of a thoracic syrinx extending from T₃ to T₉ level and arachnoiditis with atrophy of the spinal cord between T₁–T₂. During the subsequent two years, she was kept on conservative treatment while her complaint continued. Four years after having TB meningitis she encountered a progressive weakness in both legs accompanied with loss of bladder and bowel control, and numbness below the umbilicus. On re-admission (October 1996) she was found to be fully alert, conscious, well oriented and cranial nerves and upper limbs were free from the disease. Neurological examination revealed spastic paraparesis with diffuse muscular wasting. Deep tendon reflexes were brisk in both legs and Babinski’s sign was positive, abdominal reflexes were absent. Decreased sensation to pinprick and caloric stimulation with loss of position and vibration sense was found below T₆ dermatome. She had an autonomous bladder. MR examination (September 1996) of the dorsal spinal cord showed extensive arachnoiditis with multiloculated syrinxes extending from T₃ to T₁₀ (Fig. 1a,b). As compared to the previous

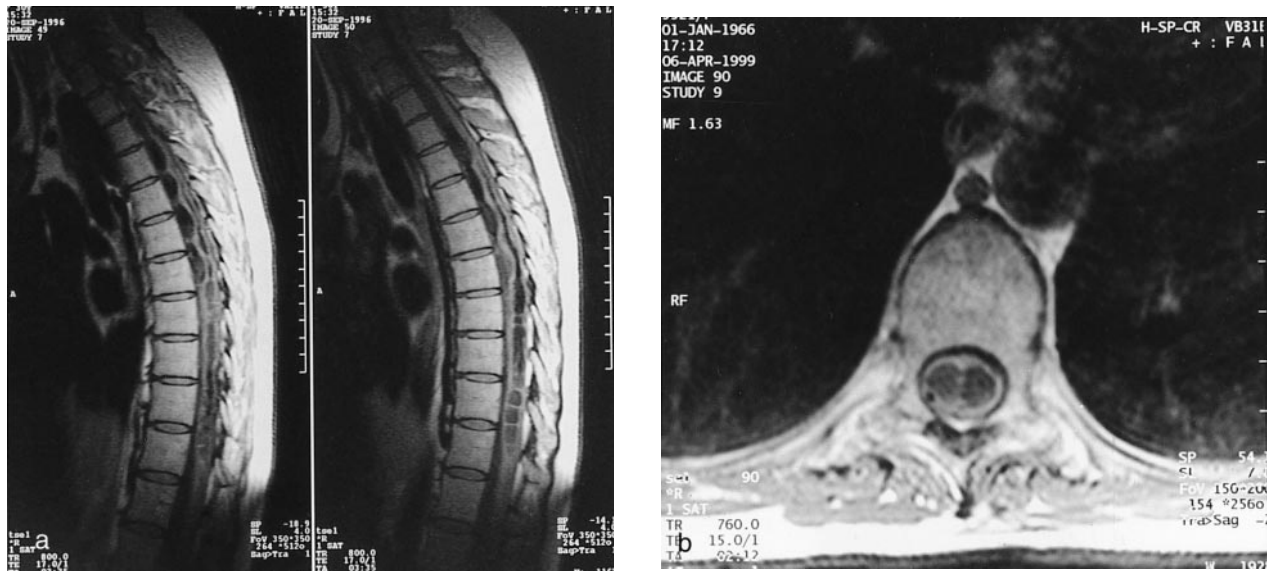


Fig. 1. On T1W (TR 800, TE 17) sagittal (a), and (TR 735, TE 15) axial (b) MRI scan of the thoracic spine showed a syrinx cavity extending from T₃ to T₁₀. Band like structures separates the cavity and attaches the spinal cord to the dura



Fig. 2. On T1W (TR 800, TE 17) sagittal (A), after shunting procedure the syrinx cavity is obviously diminished. Postoperative changes at the laminar arch and subcutaneous tissue are also observed



Fig. 3. On T1W (TR 600, TE 12) sagittal image shows re-expanding syrinx almost at the same levels

MRI, syringomyelic cavitations were found to be increased in size. These dilations of the syrinxes were thought to be the cause of the neurological deterioration, and a syringo-subarachnoidostomy was planned. On the 1st of November, left T_{5/6} hemilaminectomy was performed. After opening the dura, a thickened, cloudy arachnoid was seen. It was easily dissected from the dura, due to patchy adhesions to the spinal cord preparation of the myelotomy site was difficult. A "T" tube (Medtronic PS Medical, CA, USA) was inserted into the syrinx cavity via myelotomy. Clear fluid under moderately elevated pressure drained freely and the caudal end of the catheter was placed under the arachnoid membrane. On the first postoperative day she showed a dramatic improvement, and she managed to

walk with minimal assistance within a week. She was then referred to a rehabilitation facility. Unfortunately, this improvement progressively declined after the first postoperative month. Two years after surgical intervention, the control MRI showed that syringostomy is patent, the cavity is diminished but there seems to be no objective recovery of her neurological function (Fig. 2). Two and a half years after the operation the patient complained of having a burning type of central pain, and further deterioration in neurological function. Thoracic spinal MRI examination demonstrated enlarged syringomyelic cavities (Fig. 3). In the second operation syringo-peritoneal shunt insertion was performed via right T₁₀₋₁₁ hemilaminectomy using a "T" tube. On the first postoperative day the



Fig. 4. On T1W (TR 600, TE 12) sagittal image shows, after the second surgical procedure (syringo-peritoneal shunting) syrinx cavity is diminished once more

Table 1. Reported Cases of Syringomyelia Following Tuberculous Meningitis

Reference	Case no	Age at TBM	Onset of syringomyelia following TBM
Appelby [1]	1	19 yrs	11 yrs
	2	13 yrs	1 yr.
Barnett [3]	3	15 yrs	22 yrs
Gimenez-Roldan [9]	4	infancy	20 yrs
	5	18 yrs	4 yrs
	6	26 yrs	6 yrs
	7	10 yrs	8 yrs
	8	24 yrs	15 yrs
Savoriada [15]	9	35 yrs	11 yrs
Pépin [13]	10	12 yrs	24 yrs
	11	35 yrs	16 yrs
	12	23 yrs	28 yrs
Suzuki [18]	13,14	NA	NA
Tsuchiya [19]	15-16	NA	NA
Caplan [4]	17	40 yrs	7 yrs
Williams [20]	18	16 yrs	4 yrs
Schon-Bowler [17]	19	40 yrs	1 yrs
Fehling-Bernstein [8]	20	23 yrs	5 months
Daif [6]	21	35 yrs	6 weeks
	22	32 yrs	11 days
Kakar [11]	23	43 yrs	1 yr.
Kaynar	24	26 yrs	2 yrs

TBM Tuberculous meningitis; NA information not available.

pain was relieved, the strength of the legs improved, and the spasticity of the lower extremities decreased. The patient started an ongoing rehabilitation program three days after the operation. Ten weeks after the insertion, the control MRI revealed significantly diminished syringomyelic cavities (Fig. 4). At present, 4 months after the second operation, the patient's neurological examination dem-

onstrated decreased spasticity, and improved strength in the legs compared with the preoperative level.

Discussion

The most common cause of TB myelopathy is Pott's paraplegia [8] followed by intra or extradural granulomas, necrotising granulomatous arachnoiditis with compressive and inflammatory thrombosis of spinal cord vessels. Late neurological complications include hydrocephalus, cranial nerve palsies, vascular occlusion, spinal cord and root damage [15]. Syringomyelia is usually a late complication of tuberculous meningitis and there have been a few reports on this subject [1, 3, 4, 6, 8, 9, 11, 13, 14, 15, 17, 18, 19, 20].

Abnormal cavitation of spinal cord, called syringomyelia, was first described in the nineteenth century by Vulpian (1861) and Charchot and Joffery (1869). The first probable "TB caused" syringomyelia was described by Marinesses in 1916; a patient with chronic hypertrophied meningitis and spinal cord cavities who died of pulmonary TB. Afterwards other cases with syringomyelia secondary to TB meningitis appeared in medical journals (Table 1). The mechanism of syrinx formation due to inflammatory arachnoiditis includes ischemic myelomalacia secondary to inflammatory occlusion of spinal cord vessels with subsequent syrinx formation. Focal scarring causes a block in the circulation of CSF, thus forcing CSF into the central canal of the spinal cord via Virchow-Robin spaces. Obstruction of Virchow-Robin spaces also emerges, focal cystic dilatations in the cord eventually coalesce to form a syrinx [8]. A different possible mechanism described by Savoirda was a spinal cord fixed by arachnoidal adhesions. The cord can be lengthened due to neck movement. This action squeezes the necrotic pulp or the fluid filling the initial cavity, provoking a disruption of the spinal cord along the least resistant part. This causes upward extension of the syrinx [11, 15]. Opening of the syrinx into the subarachnoid space is a rare event but could occur in severe cases [9]. In trauma patients, it is proposed that the presence of arachnoiditis at the level of trauma causes spinal cord tethering and plays an important role in the pathophysiology of progressive post-traumatic myelomalacic myelopathy [7, 12]. Spinal cord tethering alone may lead to progressive neurological deficits despite a decompressed subarachnoid space and a collapsed syrinx [16]. However following a meningitic event arachnoid scarring is generally much more extensive.

The latent period between the initial inflammatory events and the development of symptoms related to syringomyelia is usually long and varies between 7–28 years [11, 13]. Formation of syrinx in acute stages is described by Daif *et al.* and occurred in a relatively short period of time (5 months) in Kakar's case [6, 11]. The cause of rapidly progressive myelopathy in TB has been attributed to vascular thrombosis of the spinal cord vessels [11], which was observed as an operative finding in cases in the acute stage [8]. The first clinical suggestion of enlargement of a syringomyelic cavity is usually progression of and the onset of new symptoms and signs after a stable period [2, 5].

MRI has a potential role in detecting myelopathy. Using different sequences MRI can differentiate myelomalacia and syrinx. T1W, IR, and PD images are very useful for diagnosing cystic areas due to the advent of MRI by which, TB myelopathy, syringomyelia, which are surgically treatable, can be detected in its early phase.

Untreated syringomyelia is compatible with long survival without progression in 35–50% of cases. The indication for surgical treatment either in the form myelotomy and syringostomy or shunt procedure is acute progression of neurological findings. We suggest that in postmeningitic syringomyelia a syringo-peritoneal shunt should be the initial choice, and despite long duration of neurological deficits every attempt should be made to alleviate a treatable cause of the neurological deficits. We think that the early improvement seen just after the operation is probably due to drainage of the syringomyelic cavity during the myelotomy. If the shunt continues to drain the cavity then the neurological condition may remain stable. As we have seen after the syringo-subarachnoid shunt insertion, if the shunt cannot drain sufficient amount of liquor from the cavity then the neurological status begins to worsen after a short period of time, it was 1 month in our case. Maybe it is still early to come to a conclusion about it, but 4 months after the insertion of the syringo-peritoneal shunt, the improved neurological condition was maintained. It is important to keep in mind that in such patients formation of syringomyelic cavities may be responsible for late neurological deterioration.

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Comment

Kaynar *et al.* report on a severe form of postmeningitic syringomyelia which has frustrated almost anybody who had to treat such a patient. Syringomyelia develops as a consequence of impaired CSF flow in the postmeningitic spinal subarachnoid space. The extensive arachnoiditis in such a patient makes it impossible and hazardous to attempt to untether the cord and to provide a free passage of CSF.

Thus, the cause of syringomyelia in such a case cannot be successfully treated. This leaves two options: conservative treatment or a drainage procedure. Drainage from the syrinx towards the subarachnoid space is bound to fail due to the severe disturbance of CSF flow, problems of syrinx fluid absorption, the tendency of arachnoid scarring obstructing the tube etc.. This is illustrated by the unsatisfactory result of the first operation in this case. Shunting towards an extrathecal compartment – peritoneal or thoracic cavity – offers better chances. Some authors even suggest that the peritoneal shunting is sufficient and that drainage of the syrinx, which requires a myelotomy and acceptance the risk of further neurological deficits, is not necessary (Vengsarkar *et al.* 1991).

Finally, we have to consider that the arachnoiditis itself may be responsible for progressive neurological symptoms and signs. The clinical course did not correlate absolutely with the size of the syrinx in this case. Syringomyelia is a secondary manifestation of extensive

arachnoiditis and probably not the pathophysiological component, which will determine the future prognosis in this patient. A patient has to be informed about this before the decision for a drainage procedure is made.

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