

Syringomyelia and arachnoiditis

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Abstract

Five patients with chronic arachnoiditis and syringomyelia were studied. Three patients had early life meningitis and developed symptoms of syringomyelia eight, 21, and 23 years after the acute infection. One patient had a spinal dural thoracic AVM and developed a thoracic syrinx 11 years after spinal subarachnoid haemorrhage and five years after surgery on the AVM. A fifth patient had tuberculous meningitis with transient spinal cord dysfunction followed by development of a lumbar syrinx seven years later. Arachnoiditis can cause syrinx formation by obliterating the spinal vasculature causing ischaemia. Small cystic regions of myelomalacia coalesce to form cavities. In other patients, central cord ischaemia mimics syringomyelia but no cavitation is present. Scar formation with spinal block leads to altered dynamics of cerebrospinal fluid (CSF) flow and contributes to the formation of spinal cord cystic cavities

Symptomatic or secondary syringomyelia refers to spinal cord cysts, often long and tubular, that are caused by other recognised disease processes. Intramedullary spinal cord tumours are sometimes associated with a syrinx usually because of secretion of fluids by tumour cells or necrotic cavitation of the neoplasm. Scarring of the spinal meninges (usually termed "arachnoiditis" though both the pia mater and arachnoid are involved), has also been reported as a cause of secondary syringomyelia. The arachnoiditis may be caused by trauma,¹⁻¹² pyogenic infection,¹³⁻¹⁴ tuberculous meningitis,¹⁴⁻²⁰ luetic meningitis,²⁻¹⁵⁻²¹ bleeding into the meninges¹⁸⁻²² and post operative scarring.¹³ Though arachnoiditis is known to be associated with syrinx formation, the mechanism of syringomyelia is not well understood. We report five patients with non-traumatic arachnoiditis who later developed syringomyelia.

Case reports

Case 1. This man was observed extensively during many years and studied at necropsy. In 1914 (aged 17), whilst in the army, he suffered an acute infectious illness that was diagnosed by lumbar puncture as meningitis. He made a good recovery except for temporary strabismus and continued to serve in the army until 1919. In 1935 (aged 38), he noted loss of sensation

and weakness in his right hand. Months later, he developed sensory loss and weakness of the hands, more noticeable in the left than the right. Atrophy, frequent burns, and hand tremor were reported by the patient. Four years later, the lower extremities became weak, initially on the right side. He then experienced weakness in the left leg which soon became more severe than in the right. He complained of a drawing, burning pain in both arms from the hands to the radial forearms and from the hips to the toes.

In 1943 (aged 46), he was first evaluated at the Harvard Neurological Unit at Boston City Hospital. There was diminished body hair and burn scars on his fingers. Mental function and cranial nerves were normal except for a slight left lower facial droop and slight leftward protrusion of the tongue. Upper extremity muscles, especially the deltoids and intrinsic hand muscles, were atrophied and occasional fasciculations were noted. There was bilateral lower extremity weakness with a left foot drop and atrophy of the left tibialis anticus. An inconstant intermittent "piano playing" tremor was seen in the hands. Upper extremity reflexes were absent except for a weak right triceps jerk but lower extremity reflexes were exaggerated with right ankle clonus and extensor plantar responses. Pin and thermal sensation were lost in the hands, radial forearms and entire left leg. Touch was also decreased in these parts but to a lesser degree. Vibration sense was lost at both wrists, the right elbow, and the left leg below the knee. Position sense was diminished below T3. Gait was unsteady and wide-based and was characterised by overstepping due to a left foot drop. Lumbar puncture revealed normal fluid and dynamics.

He was placed in a chronic care unit, at the Long Island Hospital, where he was examined by successive generations of Harvard Medical School students, residents and attending physicians and considered to be an example of "classic syringomyelia". He became progressively quadriplegic and bedridden and was readmitted to the Neurological Unit in 1966. At that time his mentation and cranial nerve functions were normal. There was diffuse muscle weakness; he could not lift his head from the pillow, extend his outstretched hands, sit up from a supine position, maintain wrist flexion or extension, or move his hands. The right lower extremity was completely immobile, but he could lift his left leg from the bed. The lower extremities were very spastic and sensory stimuli evoked flexor spasms in the left foot. There were no upper extremity reflexes; knee

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and ankle reflexes were exaggerated and both ankles showed sustained clonus. Both plantar responses were extensor. Position and vibration sense were impaired below C5. There was complete sensory loss to all modalities below T1 on the left and a relative loss to pin prick below T6 on the right. All sensory modalities were lost in the C5, C6 and C7 dermatomes on the right. CSF protein was 260 mg per cent, later 185 mg per cent. CSF dynamics were abnormal and myelography confirmed an irregular high cervical block. He died of pneumonia after a brief stay in another extended care unit.

At necropsy, all layers of the meninges were severely scarred and adherent to each other. The thickened dura mater was densely adherent to the underlying bony skeleton, making it difficult to extract the spinal cord intact. The meninges of the posterior fossa were also scarred. There was no cavitation or slit within the medulla. The spinal cord was thin, atrophic, and flattened, especially in the cervical and thoracic regions, but extending through lumbar segments. Microscopic sections confirmed severe fibrosis and thickening of the meninges. Both arteries and veins showed fibrous degeneration of vessel walls. At times, in the areas of necrosis, there was extensive connective tissue proliferation with accumulation of dense eosinophilic staining collagen surrounding thickened vessels. There were few remaining inflammatory cells and no granulomas.

The cervical cord was reduced to a thin ribbon with extensive central necrosis and cavity formation. The cavity was collapsed, smooth and often bilobed, but there were no lining cells nor a well formed wall. There were few remaining anterior horn cells in the cervical or upper thoracic cord. The cervical roots were thin. The dorsal and ventral roots also showed moderate to marked loss of myelin. There was some Schwann cell proliferation.

The thoracic cord was also flattened. The central zones stained poorly for myelin and had occasional small cavities. Caps of peripheral myelin remained. The corticospinal tracts were decimated throughout the thoracic and lumbar cord. The spinocerebellar tracts and posterior columns were scarred especially at the C7 and T1 region as part of the general destruction. The lumbar roots were also thin and enmeshed within adhesive arachnoidal scarring which extended to the filum terminale and cauda equina.

Case 2 In 1970, aged 36, this woman suddenly felt unwell and by the end of the day had difficulty in walking, especially with her right leg. She was admitted to hospital. The spinal fluid was bloody. She did not recall the initial two weeks in hospital, and remembered no pain, headache, or backache. After recovery, her right foot remained numb. In 1976, she noticed increased numbness in her right leg, dragging of both feet especially the right foot and urinary hesitancy. There were no acute attacks or pain. Neurosurgical exploration showed a dural arteriovenous malformation centred around the T6 level. The malformation

was removed but there were no changes in the neurological symptoms and signs in the post operative period.

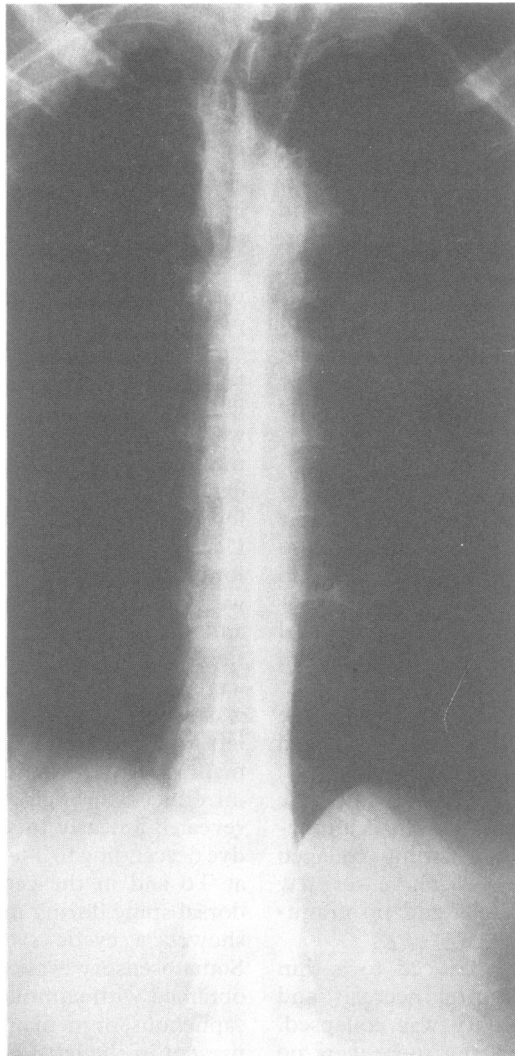
In 1981 (aged 47), she noticed increased weakness of the left leg. Urinary retention developed and was treated with an indwelling catheter. Her left hand became weak and the inside of the hand was numb. The left half of the face also became numb and she experienced occasional vertigo. Examination revealed rotatory nystagmus greater on left gaze. The pupils were small but reacted to light. Pin and thermal sensations were diminished on the left face while touch perception was normal. The left corneal reflex was diminished. There was wasting and weakness of the left intrinsic hand muscles and some weakness of the right wrist extensors and intrinsic hand muscles. The lower extremities were quite weak and spastic. Deep tendon reflexes were exaggerated except for the left biceps and brachioradial jerks which were absent. There was bilateral ankle clonus and plantar responses were extensor. Vibration, position, pain and temperature sensation were diminished below T7 bilaterally. There was decreased pin and thermal sensation in the left arm, face and neck. Cranial CT was normal. Myelography using metrizamide that we introduced through a lateral C1–C2 puncture revealed a nearly total block at T5 with some dye descending to T6–7. The cord was widened at T6 and in the cervical region. CT of the dorsal spine during metrizamide myelography showed a cystic swelling within the cord. Somatosensory evoked responses could not be obtained with stimulation of the left peroneal, saphenous, or median nerves. Denervation was present in the left C8 and T1 root innervated muscles. Blink response recording was abnormal in the left supraorbital region. The electrophysiological abnormalities were consistent with a lesion affecting the left cervical spinal cord and lower medulla. Spinal angiography was normal.

At surgical exploration, there were dense dural and arachnoid adhesions at T6, but no residual AVM. The spinal cord was dilated above the cicatrix. Incision of the dilated cord led to fluid drainage from a syrinx and a catheter was placed within the cavity. Microscopic examination of material removed at the T6 level revealed only dense fibrocollagenous tissue. Postoperatively, her legs were slightly weaker.

Two years later, she noticed increasing weakness in both hands. The left pupil was miotic and left nystagmus and left facial hypalgesia were unchanged. The left intrinsic hand muscles were weak and atrophic. She was confined to a wheelchair.

Case 3 In 1975, aged 40, this patient developed fever, myalgia and lethargy, complaints which persisted for five weeks before she was admitted to hospital with a stiff neck and stupor. Lumbar puncture contained 3,024 WBC/mm³ all lymphocytes. Protein was 90 mg/dL, glucose 23 mg/dL. After isoniazid, ethambutol and corticosteroids, she improved but complained of occasional paresthesiae below the mid thoracic level particularly on the left. She

Figure 1 Myelography (Case 3). Widened lower thoracic cord.



had urinary retention which required catheterisation. There was moderate weakness of both legs with bilateral partial foot drop, exaggerated reflexes and bilateral extensor plantar responses. Hypalgesia was present below T9. All CSF cultures were normal, but she gave a history of tuberculosis at the age of seven.

She made an excellent recovery and returned to work, but later recalled slight numbness below the waist as a residue of her meningitis. In 1982, she developed difficulty in walking and more numbness in her toes. Pain developed in her left chest and the left side below the waist felt heavy as if it were "bursting". Examination revealed atrophy of the calf and intrinsic muscles on the left. The left psoas and hamstring muscles were moderately weak and there was a severe left foot drop. The right lower extremity muscles were slightly weak. The left knee and ankle jerks were absent, but right lower extremity reflexes were exaggerated. Abdominal reflexes were absent. Spontaneous flexor spasms were present bilaterally, but the toes were immobile on plantar stimulation. Pin and thermal sensation were diminished in the feet. Walking deteriorated considerably during the following couple of months. Chest radiographs revealed bilateral apical scarring. Myelography showed a high grade block in the T4-T8 region with irregular filling of the subarachnoid space and an increased diameter

of the lower thoracic spinal cord (fig 1). CT of the dorsal region after metrizamide revealed contrast material within the cord from T8 to T11 (fig 2). The cord was widened from T7 to L1. MRI axial sections from a first generation scanner showed a large fluid compartment within the lower thoracic cord surrounded by a thin rim of spinal tissue. During the ensuing weeks, she began to notice slight tingling and prickling in her left arm and an unusual feeling around her lips and nose. There was pain in the left shoulder.

Exploration confirmed severe arachnoiditis in the region of the conus medullaris. Rostral to the scarring was a fluid filled cavity into which a silastic tube was placed and drained into the peritoneum. Postoperatively, her walking improved, but she had more difficulty initiating urination. The left lower extremity below L1 was more "numb" but the facial and arm paresthesiae were gone.

Case 4 This was a 26 year old woman, who noticed a right foot drop in 1979 followed by weakness of the right arm. There was no pain and she had no sensory complaints. The right sided weakness remained unchanged until 1981, but increased after the birth of her second child after which weakness progressed to involve the left hand. She had been admitted to hospital for meningitis at the age of one year. At that time, a gram stain of CSF showed diplococci, and penicillin was given.

Examination in August, 1983 revealed a woman of below average intelligence. Horizontal and vertical nystagmus were present and both corneal reflexes were diminished. There was slight right facial weakness and decreased hearing in the left ear. Both hands showed atrophy of intrinsic muscles. Both arms were severely weakened and the lower extremities were moderately weak. Biceps and brachioradial reflexes were absent. Lower extremity reflexes were exaggerated more on the left. The right plantar response was extensor, the left equivocal. Hypalgesia was present in both arms and in a mantle-type distribution from C6 to T2. Position sense was impaired in both hands but was intact in the legs. CSF showed 4 WBC/mm³ and normal chemistry. Skull and cervical spine films were normal with

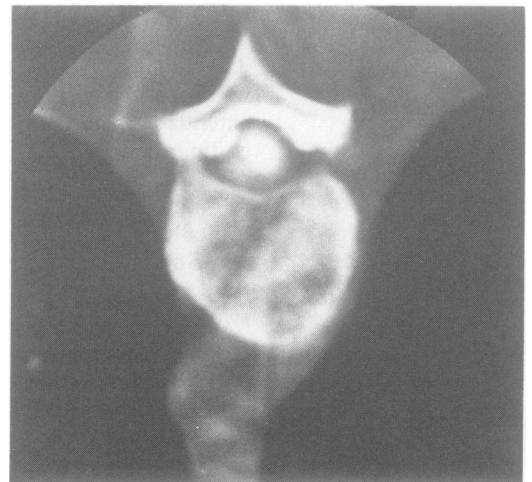


Figure 2 CT with metrizamide (Case 3). Contrast within cyst in lower thoracic cord.

no signs of basilar impression. Brain CT was normal. Myelography demonstrated meningeal irregularities and fusiform expansion of the cord from C2 to T2. CT with metrizamide documented a syrinx in the bulb and cervical cord.

Case 5 In 1946 a 58 year old man was treated for meningitis at Cook County Hospital, but the records of that admission could not be found. In 1954, his back was burned before he realised his clothes were on fire. At that time, examination revealed muscular atrophy in both arms, weakness of the arms and the right leg, spasticity of the lower limbs, absent arm reflexes and equivocal plantar responses. Pain and temperature sensibility were lost from C4 to T12 on the left and C4 to L1 on the right. Vibration sense was reduced in both hands and feet. Chest radiographs showed a calcified granuloma in the right apex. Myelography showed widening of the cervical cord consistent with syringomyelia. CSF was under normal pressure and contained 5 WBC/mm³, protein 22 mg/dL, glucose 77 mg/dL. The cervical and thoracic cord were irradiated.

In 1958, weakness increased in the arms and legs. Position sense was now impaired in the fingers and there was increased lower extremity spasticity with clonus and Babinski signs. His condition stabilised until 1979 when he was re-examined because of ataxia and increased weakness. Sensation was now decreased over the right face. Cranial CT showed dilatation of both lateral ventricles and slight dilatation of the third ventricle. The fourth ventricle was normal and there was no evidence of an Arnold-Chiari malformation. In 1981, the CT was unchanged and myelography revealed no block and an atrophic appearing cervical cord.

Discussion

Our case material includes three patients with early life meningitis that later developed symptomatic syringomyelia and syringobulbia. There was a long delay, eight, 21 and 23 years, between meningitis and spinal cord symptoms. Each had a progressive course. In two patients, CT documented a cervical cord cavity. In the third, necropsy revealed severe arachnoidal scarring with necrosis of the central portion of the spinal cord and cavity formation. All three of these patients had bulbar symptoms and signs. A fourth patient, Case 3, developed focal thoracic cord symptoms and signs with a sensory level during the acute phase of presumed tuberculous meningitis. Seven years later, she had progressive dysfunction of the spinal cord, and CT, MRI, and surgery documented a cavity within the thoracic and lumbar spinal cord. A fifth patient, Case 2, developed cervical syringomyelia and likely syringobulbia after spinal subarachnoid haemorrhage and surgical obliteration of a thoracic spinal AVM. In all patients, spinal cavities developed after meningeal scarring; in three patients, the meningeal scarring was diffuse and in two quite focal. None of the patients had evidence of Chiari-type malformation or basilar impression.

Arachnoiditis and the history of its association with syringomyelia

Extensive scarring of the spinal meninges usually involves all layers of the meninges, including the pia mater and dura mater. Nevertheless, spinal meningeal scarring is traditionally referred to as "arachnoiditis" especially when the scarring causes loculation and interrupts and distorts the flow of contrast media introduced during myelography. The meningeal spaces, the subdural and subarachnoid compartments, usually communicate and inflammatory disorders usually affect each space to some degree.²³ Spinal arachnoiditis may be quite focal, and was originally described as "arachnoiditis adhesiva circumscripta spinalis" or generalised, "arachnoiditis adhesiva diffusa spinalis".

Vulpian (1861) and Charcot and Joffrey²⁴ were probably the first to describe cervical spinal cord cavitation associated with arachnoiditis found at necropsy. Schwarz²⁵ commented on chronic arachnoidal scarring in a patient with syphilitic sclerotic and cystic meninges who had focal regions of softening and cavitation within the spinal cord. Alajouanine *et al*²⁶ in a presentation to the Paris Neurological Society, described six patients with spinal arachnoiditis especially affecting the posterior aspect of the meninges who also had syringomyelia. Mackay²⁷ in a report on chronic spinal arachnoiditis reviewed the literature and reported five of his own patients defining the clinical and pathological features of chronic adhesive arachnoiditis. At necropsy two of Mackay's patients showed severe arachnoiditis with intense thickening of vessel walls in the meninges. Both patients had spinal cord softening, but cavitation was present in only one patient. Mackay pointed out that "cavitation of the cord may occur as a result of chronic arachnoiditis and future studies on "syringomyelia" must take this into account". Barnett¹³ analysed previous reports, added new cases, and summarised the then current information about chronic arachnoiditis and its association with syringomyelia. Three different patterns of arachnoiditis have been associated with syringomyelia: focal or diffuse scarring of the basal posterior fossa meninges, diffuse adhesive spinal arachnoiditis and focal spinal meningeal cicatrix formation.²⁸⁻³⁰ Focal scarring and opacification of the ventral surface of the brainstem has been associated with focal cystic lesions in the midbrain, communicating with the cerebral aqueduct, the "keyhole aqueduct syndrome" a special form of syringomyelia.³¹

Basal arachnoiditis

Scarring of the basal meninges in the posterior fossa is a recognised precursor of syringomyelia, but we had no examples of this condition in our data. Newman *et al*²⁸ proposed that birth trauma, unusually frequent in the history of patients who later develop syringomyelia, is important in causing posterior fossa arachnoid scarring in both normal infants and those with Chiari malformations. McLaurin *et al*³² produced cervical

spinal cord cavitation experimentally in dogs by injecting kaolin into the cisterna magna. As with other posterior fossa and foramen magnum anomalies and lesions, intracranial pressure may be transmitted to the cerebral spinal cord causing a "communicating syringomyelia".³³ Gardner³⁴ proposed that CSF travelled down into the dilated central spinal canal from the floor of the fourth ventricle. Blockage of free flow of CSF within the subarachnoid space at or above the foramen magnum must somehow facilitate syrinx formation.

Trauma with delayed syringomyelia

Focal scarring of the meninges has been known to cause syrinx formation especially in patients who have had spinal trauma. Gordon Holmes¹ in his Goulstonian lectures on spinal cord injuries commented on "curious cavities, roughly cylindrical . . . in the segment of the spinal cord adjoining the lesion either in those above it or below or both." These often extended a considerable distance, in some cases four or five segments and at the ends furthest from the wound they were occasionally the only indication of any abnormality. Some cavities were found above and below the lesion while others occurred just below it. Barnett *et al*¹⁵ studied 591 patients with traumatic paraplegia seen during an 18 year period and identified eight who later developed spinal cord symptoms rostral to their original traumatic lesion. Four of these eight patients had myelographic confirmation of cervical spinal cord enlargement. Upper extremity paresthesiae, numbness, weakness, or reflex loss and sometimes facial numbness developed one to 14 years after the original trauma (average five to six years). Progressive late post-traumatic syringomyelia was identified in six of 659 patients (0.9 per cent) admitted to a referral centre for spinal cord injuries.⁴ Syringomyelia was diagnosed five to 23 years (average 13 years) after the original traumatic injury. Shannon *et al*⁵ reported 13 patients with post-traumatic syringomyelia. Incomplete spinal cord lesions were as commonly complicated by late syrinx formation as were patients with complete spinal cord transections.

In patients with post-traumatic syringomyelia, the initial symptom referable to the syrinx is usually pain, especially precipitated by coughing, sneezing, or otherwise straining; paresthesiae, numbness, and unwitting hand injuries and burns follow and paresis and atrophy develop much later.³⁻⁹ Sensory symptoms can involve the face as well as the upper limbs. When syringo-peritoneal shunting of CSF has been helpful, the relief of pain has been the first and most frequent symptom to show improvement.^{20 35} At surgery or necropsy in patients with a previous trauma, there is usually scarring of the meninges with cicatrix formation at the site of maximum injury as well as a syrinx. Occasionally, a large accumulation of fluid forms within the meninges as an arachnoid cyst that compresses the spinal cord externally.¹³

Theories abound as to why the syringomyelia cysts form and why they expand. A

traumatic intraspinal haematoma could develop at the time of the injury and later become hydrolysed leaving a slit-like cavity. Alternately, the cord could be torn or sheared during the injury, leaving a myelomalacic cord. This softened area would be in continuity with the subarachnoid space immediately after trauma. Gliosis develops at the edge of the tears so that the slit-like cavities are lined by glial scars. Mechanical factors might force spinal fluid up or down the cavities thus expanding them.^{10 34} Once the cavity has begun to form, subsequent cranial-spinal pressure differences,³⁴ "sloshing" of fluid³³ or mechanical forces related to respiration, movement, or straining⁷ could cause gradual enlargement of the cavities. These hypotheses do not explain the sometimes long delay in symptoms; gradual enlargement of an intramedullary lesion caused by an intraspinal slit or haematoma should be associated with severe paraplegia directly after injury, and progression should begin soon after. Alternatively, the mechanism of late syrinx formation could involve the meningeal cicatrix not just the intraspinal lesions. Meningeal fibrosis and scarring with adhesions takes time to develop.

Trauma is an excellent example of focal meningeal scarring (meningitis circumscripta). Two of our patients developed syringomyelia after focal non-traumatic scarring. One patient had a spinal AVM that bled and was repaired surgically. Later exploration confirmed a severe focal adhesive meningeal scar. The second patient had tuberculous meningitis with focal signs in the thoracic cord which cleared rapidly. The patient with focal bleeding developed worsening of function in spinal segments above and below her AVM scar five years after surgery, and the patient with tuberculous meningitis deteriorated seven years after her acute infection and a syrinx developed below her prior lesion. Both patients had cord injury as well as meningeal inflammation. They are thus comparable to patients with trauma and incomplete paraplegia.

Non-traumatic spinal arachnoiditis and syringomyelia

Barnett¹³ reviewed seven well documented previous examples of syringomyelia due to non-traumatic spinal arachnoiditis and added five new cases. Among these twelve cases antecedents were: subarachnoid haemorrhage,² pyogenic meningitis,² Pott's disease,¹ spinal anaesthetic,¹ spinal surgery,² and unknown.⁴ In some patients, spinal cord dysfunction was recognised at the time of the injury or illness, but in other patients symptoms were delayed for as long as 13 to 17 years (average delay 4.6 years). The cavities in the patients in Barnett's series¹³ always involved the spinal cord maximally in the segments beneath the thickened meninges and always affected the thoracic cord. In nine patients, there was severe localised arachnoid scarring and three patients had cystic fluid collections within the scarred arachnoid.

Tuberculous meningitis

Tuberculous meningitis is often complicated by transverse myelitis and polyradiculitis and can cause chronic meningeal scarring, adhesions and spinal block.¹⁶ Marinesco²¹ described a patient with chronic "hypertrophic" meningitis and spinal cord cavities who died of pulmonary tuberculosis. Savoiaro¹⁷ reported a patient with tuberculous meningitis who was treated with intrathecal streptomycin and isoniazid. Slight paraparesis accompanied the original infection and, during a relapse the following year, the patient became severely paraplegic. An arachnoidal scar and complete spinal block were demonstrated. After a stable period of 12 years, he developed arm paresthesiae, weakness, and numbness. Air myelography confirmed a pocket of arachnoiditis with a block at T10 and an air-filled cavity extending rostrally to C1. Feigen *et al*¹⁸ described a patient with Pott's disease with severe kyphoscoliosis who had a segment of spinal cord removed that showed a syringomyelic cavity. Barnett¹³ also described a patient with Pott's disease who became paraplegic at the age of 15 in association with tuberculous osteomyelitis of T2, T3 and T4. She recovered but later developed a syringomyelic cavity and arachnoidal adhesions were identified at surgery. Barnett has reviewed the literature on Pott's disease and syringomyelia; most case reports were before 1920.¹⁵ Paraplegia in patients with Pott's disease has been found in patients without spinal obstruction and is usually attributed to an arteritis and cord ischaemia. Gimenez-Roldan *et al*¹⁹ added four new cases of tuberculous meningitis and delayed syrinx formation. One patient had syringobulbia, syringomyelia and hydrocephalus following severe early life tuberculous meningitis with scarring in the posterior fossa. The other three patients all developed tuberculous meningitis in their second or third decades (ages 10, 18 and 26 years) and all received intrathecal streptomycin. None had recognised spinal cord symptoms or signs during the acute infection. Signs of spinal cord dysfunction began four, six, and eight years later and progressed rapidly and inexorably thereafter. All had complete spinal block in the thoracic region with fragmentation of dye in other regions; all had cavities above the level of the block. Our single patient (Case 3) with tuberculous meningitis had symptoms at the time of meningitis that cleared but later developed a syrinx below the level of the previous symptoms.

Diffuse meningitis and syringomyelia

Pyogenic meningitis probably causes diffuse spinal and cranial meningeal inflammation. Syringomyelia has been described after pyogenic infection but most often the offending organism has not been identified.^{13 14 27} The diagnosis of pyogenic infection is made because of the extensive pleocytosis which argues against a viral meningitis. Diffuse adhesive arachnoiditis is probably more common in underdeveloped countries and is caused by a mixture of pyogenic, tuberculous, fungal and parasitic infections. Jenik *et al*¹⁴ identified 507

patients referred to their centre in Addis Ababa with clinical findings indicative of arachnoiditis. Among 200 selected cases studied, 90 per cent had dissociated sensory loss, a "syringomyelia syndrome," and 95 of 105 patients that had myelography had radiographically confirmed adhesive arachnoiditis. Some patients with diffuse meningeal inflammation also have focal regions of more severe scarring which may be associated with focal spinal myelographic block. We have reported three patients (cases 1, 4, and 5) with presumed pyogenic meningitis. All had meningitis in youth and no details were available concerning the signs and symptoms that accompanied the acute infection. One had presumed pneumococcal infection but no pathogen was identified in the other two patients. Patient one, at necropsy, had a diffuse adhesive meningitis, worse in some regions. In these three patients symptoms related to syringomyelia were delayed an average of 17 years.

Subarachnoid or spinal bleeding and syringomyelia

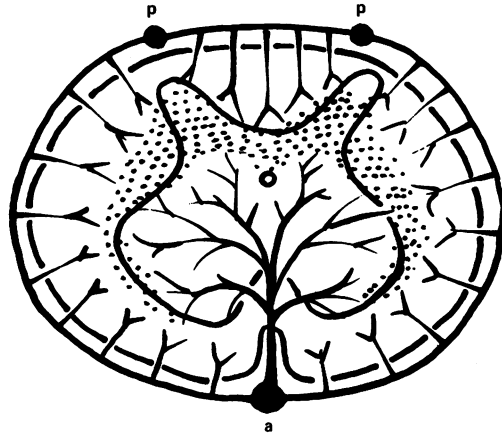
We could find only two previous reports of syringomyelia following spinal subarachnoid bleeding. A 37 year old woman with systemic lupus erythematosus developed a "thoracic transverse myelitis" with associated subarachnoid and subdural haemorrhage.¹⁸ This patient remained paraplegic until her death from active lupus. At necropsy, the dura over the lower thoracic spinal cord was firmly adherent to the leptomeninges and spinal cord and there was a large cavity at this and lower spinal levels. Blood pigment was found within the meningeal scar tissue. Nelson²² described a man with chronic thrombocytopenic purpura and a confirmed subarachnoid haemorrhage who developed a paraparesis and spinal block during a period of four years. Necropsy showed extensive thickening of the dura mater and pia-arachnoid and extensive cavitation of the thoracic and lumbar segments of the spinal cord. Our patient with a spinal AVM had an episode of subarachnoid haemorrhage but also had spinal surgery to remove the AVM. Symptoms of syringomyelia developed five years after her surgery and 11 years after the subarachnoid haemorrhage.

Conclusions and mechanisms of syrinx formation

We believe that analysis of our case material and previous cases of arachnoiditis (traumatic and non-traumatic) favours two predominant mechanisms of syrinx formation.

1. *Vascular.* The inflammatory disorder in the meninges leads to severe scarring of the meninges and the vessels that travel within the meninges. Barnett^{13 15} and Mackay²⁷ both described the presence of severe vascular changes and patient 1 showed extensive obliteration of spinal cord feeding arteries. These extensive vascular changes lead to cord ischaemia. The most severely involved segments are those supplied by the anterior spinal arteries which derive from radicular arteries each 4 or 5 segments, and in the arterial boundary zones between the anterior and posterior spinal

Figure 3 Diagram of arterial supply of cord. Border zone in the centre zone of the cord is depicted in broken lines.



arteries (fig 3). This area includes the central cord and the crossing spinothalamic fibres, and the anterior horns and corticospinal tracts. Ischaemia gives rise to areas of softening and cavity formation. This can occur focally in the case of focal meningeal scarring or multifocally when the meningeal scarring is more diffuse.

MacDonald *et al*¹² reported two patients whose findings supported a vascular mechanism of damage associated with arachnoiditis. Both patients had severe spinal trauma and developed delayed symptoms and radiological findings suggesting delayed syrinx formation. At surgery, each had arachnoidal scarring and the spinal cord contained multiple small microcysts less than 1 mm in diameter that undoubtedly represented an ischaemic myelomalacia but cysts had not yet coalesced to form true syringomyelic cavities.

A syringomyelic syndrome could occur when the central spinal structures are ischaemic even when true cavities do not form. In that case, ischaemia would mimic syringomyelia. In other cases, microcysts probably form in areas of spinal cord softening and coalesce to form true cavities. Our patient 1 with necropsy data is an excellent example of arachnoiditis, causing ischaemic myelomalacia and secondary cavitation.

2. Focal scarring with spinal block. Spinal block interferes with the circulation of CSF. Altered mechanics and pressure could force CSF into the central canal and cause expansion of that structure.³⁵ Alternatively, expansion of the central canal or cord cavity could extend into areas of damage for example, during trauma or infection. The following case supports this explanation.³⁶ A 73 year old woman developed back pain and a large syringomyelic cavity was discovered in the conus medullaris at surgery. Above the lesion was a calcified meningioma at T9 which had compressed the spinal cord altering drainage below. Examples of posterior fossa brain tumours with subsequent spinal syrinxes have also been reported.^{37,38} The mechanism of syringomyelia with Arnold-Chiari malformations and posterior fossa arachnoiditis are abnormal spinal fluid circulation and drainage. Some cerebrospinal intramedullary tumours might also produce cavitation by causing a spinal block.

The long duration of latency between the initial trauma, inflammatory event, or bleeding and the frequent absence of important spinal cord symptoms and signs at the time of the illness support the argument for the importance of the meningeal process rather than a primary intramedullary aetiology of the syrinxes. In fact, most patients had no neurological symptoms and signs in the interval between the initial injury or illness and the development of the syrinx. The initiating event sets off the process of scarring outside the spinal cord in the spinal meninges. Ultimately, the meningeal scarring becomes severe and adhesive leading to complete or nearly complete spinal myelographic block. This period of development of adhesive changes can be months, but more often is years. The spinal block interferes with CSF circulation and causes spinal cord ischaemia, leading to cavitation or multiple areas of myelomalacia. The advent of MRI and metrizamide myelography with CT may help to clarify the mechanism of this process in the future.³⁹

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Robert Graves and multiple neuritis

Many studies of paralysis of the limbs had been conducted in the 18th and 19th centuries, but the concept that this might result from diseases remote from the brain, cord and spinal roots was not recognised. Wasting was due to disorders of the muscles and sensory loss related to defects of the skin. It was recognised that these symptoms occurred in "alcoholic paraplegia" and probably in lead and arsenic poisoning where they were caused by the diarrhoea or constipation. Samuel Wilks had thought the cause might be due to a reflex paralysis "the cord is in no way structurally altered, and therefore may . . . recover . . ."¹

Polyneuropathy is a recent name replacing Ernst von Leyden's (1832-1910) term² "multiple neuritis". The first account was probably one of beriberi neuritis described by Bontius in 1642.³ Robert Graves (1796-1853) in 1843 deserves credit for first predicating disease of the peripheral nerves as a cause of paralysis. Graves, remembered for his description of exophthalmic goitre (after Caleb Parry), was a Dublin graduate who had travelled with the artist Turner in Europe. In Paris in 1828 he observed the remarkable and obscure, epidemic of acute sensori-motor polyneuropathy. Described by Auguste-François Chomel⁴ it was known as *épidémie de Paris*. Graves's account in 1843 is to be found in his *Clinical Lectures*⁵:

"One of the most remarkable examples of disease of the nervous system commencing in the extremities, and having no connection with lesions of the brain or spinal marrow, was the curious *épidémie de Paris*, which occurred in the spring of 1828. Chomel has described this epidemic in the 9th number of the *Journal Hebdomadaire*, and having witnessed it myself in the months of July and August of the same year, I can bear testimony to the ability and accuracy of his description. It began (frequently in persons of good constitution) with sensations of pricking and severe pain in the integuments of the hands and feet, accompanied by so acute a degree of sensibility, that the patients could not bear these parts to be touched by the bed-clothes. After some time, a few days, or even a few hours, a diminution, or even abolition of sensation took place in the affected members, they became incapable of distinguishing the shape, texture, or temperature of bodies, the power of motion declined, and finally they were observed to become altogether paralytic. The injury was not confined to the hands and feet alone, but, advancing with

progressive pace, extended over the whole of both extremities. Persons lay in bed powerless and helpless, and continued in this state for weeks and even months . . .

At last, at some period of the disease, motion and sensation gradually returned, and a recovery generally took place, although, in some instances, the paralysis was very capricious, vanishing and again reappearing.

The French pathologists, you may be sure, searched anxiously in the nervous centres for the cause of this strange disorder, and could find none; there was no evident lesion, functional or organic, discoverable in the brain, cerebellum or spinal marrow . . . Can anyone . . . hesitate to believe that paralysis . . . may arise from disease commencing and originating in the nervous extremities alone?"

Gowers gave him less than credit due, but Trousseau remarked that "Graves had created a class of peripheral or reflex paralyses . . ." Octave Landry's celebrated report of ascending paralysis⁶ in a 43 year old paver followed in 1859 but, curiously, the peripheral nerves were not examined.

It was left to Louis Duménil (1823-90) to demonstrate in a 71 year old stone cutter afflicted by a Landry's paralysis the *pathological evidence* of "a genuine atrophy of the medullary substance of the peripheral nerve tubes" and related loss of transverse striations of shrunken muscle fibres in the periphery.⁷

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