SYRINGOMYELIA WITH ARNOLD CHIARI I MALFORMATION: A REPORT OF FOUR CASES

S H Lim, M C Wong, K Puvan

ABSTRACT

4 cases of syringomyelia with type I Arnold Chiari malformation were seen presenting mainly with dissociated sensory loss, weakness of hands and upper motor neurone signs in the lower limbs. The first patient improved with posterior fossa decompression. The second and fourth patient appeared not to have improved after surgery, and the third case refused operation. One patient showed cord atrophy on myelogram presumably due to a collapse of the syrinx. One of the patients was unusual in that the syrinx extended down to segment T11. Magnetic resonance imaging of the posterior fossa and the cervical cord, to date, is the most useful procedure for diagnosis.

Key Words: Syringomyelia, Arnold Chiari Type I Malformation, CT scan, MRI, myelogram

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INTRODUCTION

Syringomyelia is a rare chronic spinal cord disease characterised pathologically by cavitations within the spinal cord (1). Syringomyelia associated with dilatation of the central canal of the spinal cord or hydromyelia is often associated with posterior fossa developmental anomalies such as Type I Arnold Chiari malformation (2). The latter is a condition in which the cerebellar tonsils are displaced into the upper part of the cervical canal.

We report four consecutive cases of syringomyelia associated with Type I Arnold Chiari malformation. These patients were seen at our department over a period of one year.

CASE REPORTS

Case I

A 26-year old man first noticed inability to feel the temperature of the hot bath over his left shoulder in 1982. In 1983 he had one episode of painless burn over his left forearm. Since then he developed an insidious and progressive numbness involving the left side of the neck spreading to the left upper limb, right upper limb and later to the left side of the trunk down to the middle of the left thigh. At the same time, he developed progressive weakness of both hands associated with stiffness of the lower extremities. There were no other neurological symptoms.

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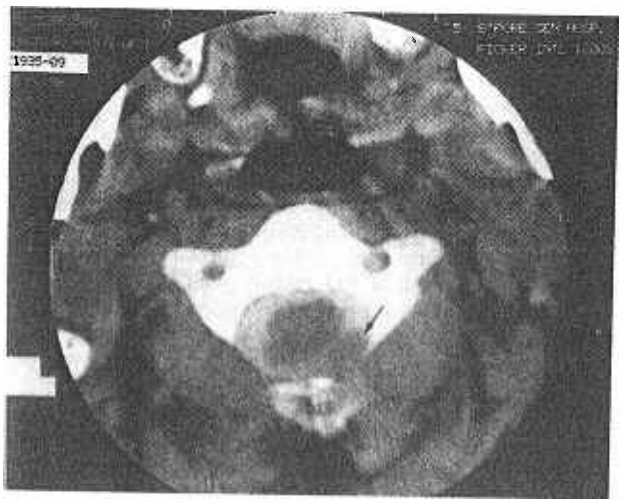

Figure 1:
CT scan of the cervical spine after cervical myelogram showed the left cerebellar tonsil extended down to the level of C2.

Clinically there was an old scald mark over left forearm. Upper limbs were hypotonic with hyporeflexia on the right but normoreflexic on the left. Weakness and wasting of the upper extremities were confined to the hands. The lower limbs were spastic with mild weakness. There was loss of pain and temperature sensation with preservation of touch, position and vibration senses over left C2 to L2 and right C2 to T7 dermatomes. The rest of the neurological examination was normal.

Cervical myelogram showed an increase in the transverse diameter of the cervical spinal cord. CT scan of the cervical spine 16 hours after cervical myelogram did not show contrast uptake in the cervical central canal. The left cerebellar tonsil extended downwards to the level of C2 (Figure 1). These findings were confirmed at operation. CT scan of the brain was normal.

Foramen magnum decompression was done. Post-operatively, his numbness had improved and the lower limb was less spastic. The rest of the neurological status remained the same.

Case II

A 40-year old woman presented with a 30-year history of progressive weakness of upper limbs which she attributed to a fall. She did not notice any numbness but scalded herself with hot objects on many occasions. There were no other significant symptoms.

Clinically she had gross wasting of the muscles of both upper limbs, especially the left. Upper limbs were hypotonic and areflexic while the lower limbs were spastic. The power in the upper limbs was generally grade 2 on the right and grade 1 on the left, affecting the flexors and extensors equally. The lower limbs showed a grade 4 weakness of the flexor muscles. There was dissociated sensory loss to pain and temperature over C4 to T1 dermatomes. The rest of the neurological examination was normal.

The cervical myelogram showed that the cervical spinal cord was slightly narrowed. CT scan of the upper cervical cord after myelogram showed a collection of contrast in a large oval shaped central canal of the cervical spinal cord. There was herniation of left cerebellar tonsil. CT scan of the brain was normal.

At operation, a swelling was seen at the level of the second and third cervical spinal cord. Both cerebellar tonsils were below the level of foramen magnum with the left tonsil slightly lower than the right. Suboccipital craniotomy, C1 to C3 laminectomy and syringotomy were performed. Postoperatively her neurological symptoms and deficits remained static after 2 years of follow up.

Case III

A 43-year old woman presented with a six-year history of progressive numbness followed by weakness involving the left hand, spreading to the whole of left upper limb and later the right upper limb. Subsequently the numbness progressed to involve the chest anteriorly and posteriorly. She had scalded herself on numerous occasions. The rest of the history was unremarkable.

The upper limbs were wasted especially distally and on the left. There were many old superficial scars. The upper limbs were hypotonic and areflexic with a power of grade 1 over 5 on the left and grade 3 over 5 on the right. The lower limbs were spastic with a power of grade 4 over 5. There was diminished sensation of all modalities including the vibration and position senses but especially the pain and temperature from left C5 to L1 dermatomes. She had a left Horner's syndrome. The rest of the examination was normal.

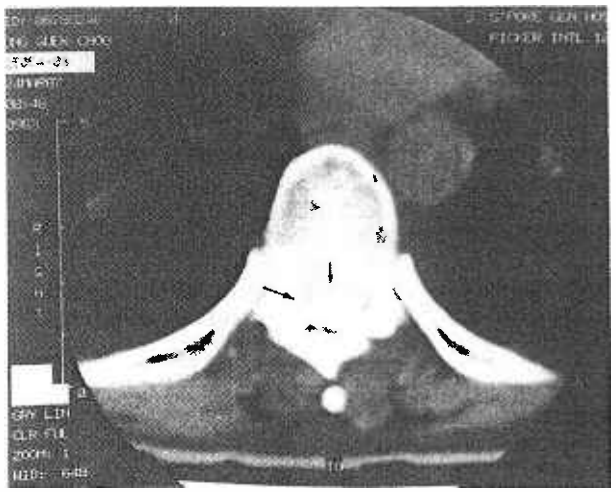

Figure 2:
Delayed CT scan of the thoracic spine after myelogram at the level of T8 showed an increase in the anterior posterior diameter of the spinal cord with a collection of contrast in the syrinx cavity (smaller arrow). There was also contrast in the subarachnoid space (bigger arrow).

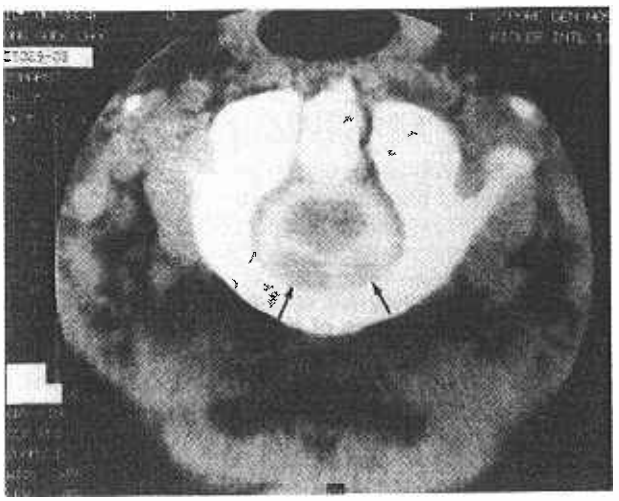

Figure 3:
CT scan of the cervical spine after myelogram at the level of C1 showed bilateral cerebellar tonsilar herniation giving rise to an inverted "Mercedes Benz" sign.

Myelogram revealed that the cervical and the thoracic spinal cord was enlarged to the level of T10 narrowing the subarachnoid space. CT scan after myelogram confirmed this enlargement. Delayed scan revealed a large spinal central canal almost occupying the entire core and extended down to T11 (Figure 2). Low lying cerebellar tonsils was seen at C1 (Figure 3).

Operation was offered but she refused.

Case IV

A 42-year old woman first noticed right sided ptosis in 1962 but did not seek any medical attention. She started having weakness and numbness in the right hand six years before consultation in 1987 . Two years later the right forearm and upper arm became involved. The left hand became similarly affected 2 years before presentation. She also noticed her upper limbs became smaller with several burns marks. She was known to have rheumatic heart disease detected in 1962. She had been asymptomatic while on diuretics and digoxin.

She had marked wasting involving the hands, forearms and arms, especially the right side. The upper limbs were hypotonic while the lower limbs were spastic. The power of both upper limbs was about grade 3 over 5 proximally and grade 2 over 5 distally. She had a mild upper motor neuron pattern of weakness in the lower extremities. There was diminished sensation to pain, temperature and touch over C8 to T1 dermatomes bilaterally. There was a right Horner's syndrome. The rest of the neurological examination was normal. Cardiovascular examination revealed mitral stenosis, mitral incompetence, aortic stenosis and aortic regurgitation.

The magnetic resonance imaging (MRI) of the posterior fossa and the cervical spine revealed an extensive syringomyelia with an associated cerebellar tonsils herniation (Figure 4 & 5). Operative findings were similar to the MRI. Suboccipital crainectomy, laminectomy of C1 to C4 and syringotomy were performed. Neurological status remained static one year after the follow up.

DISCUSSION

Syringomyelia is a chronic, often slowly progressive degenerative disease characterised pathologically by the presence of one or more cavities in the central portion of the spinal cord. It predominantly affects the cervical region but cavities may extend caudally involving the thoracic and even lumbosacral segments or cranially producing syringobulbia (3-4).

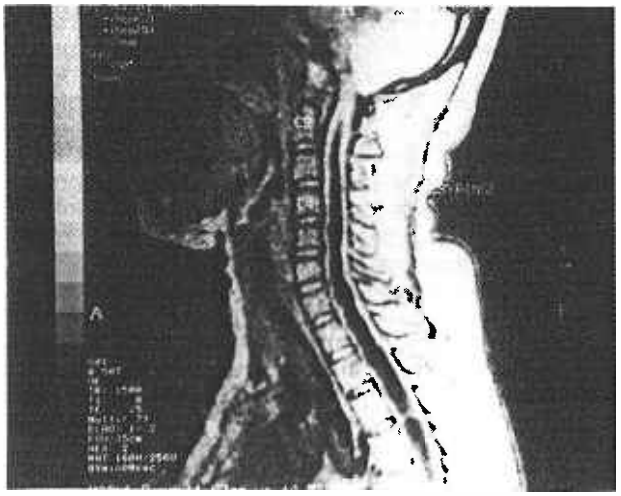

Figure 4:
MRI of the cervical & thoracic spine showing extensive syringomyelia extending from C2 down to T7.

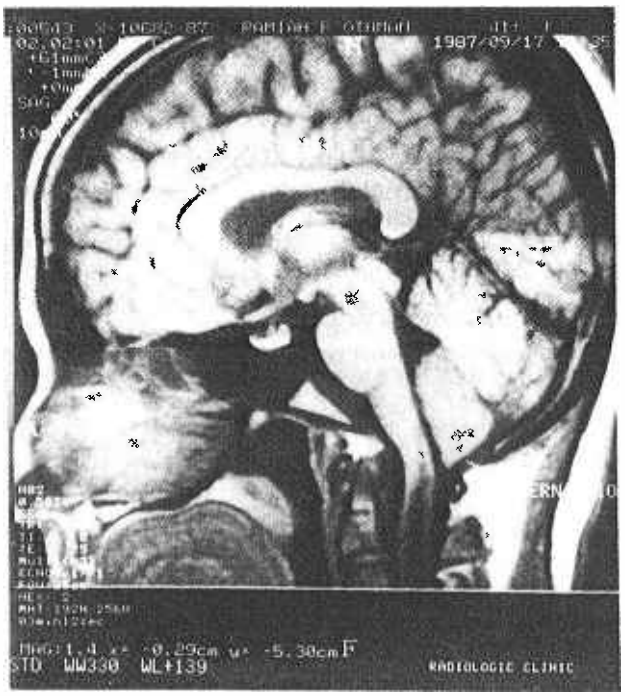

Figure 5:
MRI of the posterior fossa showing cerebellar tonsil herniation.

It is classified pathologically into two main types (2). The more common type is due to primary dilatation of the central canal of the spinal cord. The fluid within the canal transgresses through the wall, penetrating into the cord parenchyma forming irregular channels both vertically and outwards. The syrinx has communication either with the fourth ventricle or subarachnoid space, and, thus, at times referred to as communicating syringomyelia (5). This type is almost always associated with developmental anomaly, and in up to 90% of cases, Arnold Chiari I malformation. All our four patients belong to this group. The other type of syringomyelia does not have primary involvement of the central canal. The cyst arises in the cord substance without communication with the fourth ventricle or subarachnoid space. This may be due to spinal trauma, tumour or arachnoiditis.

Arnold Chiari I malformation is the first of the four categories of hind brain anomaly described by Chiari (6), and in essence comprises a herniation of the tonsils of the cerebellum through the foramen magnum with or without downward displacement of the medulla. The significance of cerebellar tonsillar position has been studied by Barkovich et al (7). They concluded that more than 2mm of tonsilar ectopia is of clinical significance especially in the presence of syringomyelia. All our patients had significant cerebellar tonsilar herniation.

The neurological symptoms and signs in syringomyelia with Chiari malformation have been extensively reviewed by many authors (1-5). Basically, they derive from a combination of 3 mechanical factors: direct compression by the tonsils of the fibre tracts and nuclei in the medulla and upper segments of the cervical cord within the foramen magnum and upper cervical canal, stretch of the cranial nerves from as high as fifth cranial nerve to the last cranial nerve, and distortion or disruption of the long tract in the spinal cord by the central cyst itself. Obviously the clinical features depend on the relative severity of each factor. All the four patients had radiological evidence of cerebellar tonsils herniation without downward displacement of the medulla. Only the first patient showed significant spinal cord compression by the cerebellar tonsil on the CT scan. Therefore the clinical features in all our patients were mainly attributed to the syringomyelia when they characteristically presented with an insidious onset and progressive dissociated sensory loss (except for the third patient when she had impairment of other sensory modalities), asymmetrical lower motor neuron lesion in the upper extremities, and lower limbs spasticity with upper motor neuron weakness. Even though the presence of posterior column signs in the third patient could be attributed to the Chiari malformation, we felt that it was the result of extensive syringomyelia when the syrinx cavity almost occupied the whole spinal cord. Two of them with thoracic spinal cord involvement had Horner's syndrome ipsilateral to the side with more severe lower motor neuron signs. None of them had lumbo-sacral involvement or syringobulbia.

Many authors have stated that the specific signs and symptoms depend on the anatomy and dimensions of the syrinx, in particular its cross sectional location and length, which determine the extent of dermatome involvement (3-4). This is true in the majority of cases and is well illustrated in our third patient when her distribution of sensory loss correlated well with the radiological findings of extensive syringomyelia. However, Grant et al did not find any significant relationship between the neurological findings and the imaging dimension of a syrinx (8). Our fourth patient illustrated their point when her sensory loss was confined only to C8 and T1 while the MRI showed extensive syrinx. This point is important to bear in mind when surgical management is being considered.

Imaging procedures are the mainstay of investigation of the syringomyelic syndrome and are directed toward confirmation of the clinical diagnosis, recognition of associated abnormalities and selection of appropriate surgical procedures. CT metrizamide myelograghy was found to be more sensitive than conventional metrizamide myelography in the diagnosis of both Arnold Chiari malformation and syringomyelia (9). It was also found that delayed (12 to 24 hours) scans demonstrated more syrinx cavities than the earlier film, as in our third patient. However the appearance and extent of the syringomyelia, and the associated high brain anomaly are best demonstrated by the MRI (10-12), as shown in our fourth patient. This non-invasive imaging method has become the first line investigation to be performed in patients whose symptoms are suggestive of syringomyelia.

The association of Chiari I malformation in all the four patients with syringomyelia is probably not by chance but play an important role in the pathogenesis of the later. According to the hydrodynamic theory postulated by Gardner (13) and later modified by Williams (14), syringomyelia probably begins with complete or partial obstructions of the cerebro-spinal fluid (CSF) outflow from the fourth ventricle into the subarachnoid space, thus leading to abnormal persistence of the central canal and its communication with the fourth ventricle via the obex. This allows the transmission of pressure waves generated by transient rises in intracranial pressure to the central spinal cord, thereby resulting in its gradual dissection and enlargement of the syrinx, which frequently occurs asymmetrically. Aboulker (4,12), however, believed that a syrinx is an alternative intramedullary route toward the absorption of CSF which is unable to follow its normal ascending course due to blockage at the foramen magnum. The CSF enters not through the upper end of the central canal, as it was shown on serial microscopic section to be totally occluded or very narrow and not capable of trasmitting fluid in a pulsatile fashion (15), but through other pathways such as Virchow-Robin spaces or the dorsal root entry zone. The CT scan after the myelogram in our third patient has lent support to this theory in demonstrating filling of the syrinx in the absence of metrizamide in the fourth ventricle.

The natural history of syringomyelia is variable and unpredictable (16). This uncertain progression of the clinical course presents considerable difficulties in the timing of surgical intervention. Untreated syringomyelia seems compatible with long survival and no progression in 35 to 50% of patients (17). Therefore surgery is best reserved for those with progressive motor deficits, which was the reason for operation in three of our patients. All of them had foramen magnum decompression alone hoping to relieve the hydrodynamic disturbance caused by the Chiari malformation. All of them benefitted from this procedure in terms of halting the progression of the disease and there was symptomatic improvement in the first patient. This outcome is expected as there was no significant spinal cord compression by the cerebellar tonsils. This is consistent with the observation that was made by Logue (2). Drainage procedure, which was not performed in all the operated patients, is reserved for those with noncommunicating syringomyelia, severe pain or hyrocephalus (18).

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