CASE REPORT

Syringomyelia with atypical presentation

AH Sarder[1], SM Kamal[2], GC Biswas[3], MS Islam[4], MA Bakar[5]

Summary

A 50 years old male patient was admitted in Khulna Medical College Hospital with grade-III weakness of both upper limbs and grade-IV weakness of both lower limbs, marked wasting of muscles in upper back and shoulder girdle region. There was no fasciculation, no pyramidal signs and no sensory loss. All routine investigations including NCS and EMG were normal. MRI of cervical and dorsal spine revealed a large syrinx extending from the cervico-medullary junction to the sixth dorsal spine with herniation of cerebellar tonsil through foramen magnum. With these findings the case was diagnosed as syringomyelia with atypical presentation.

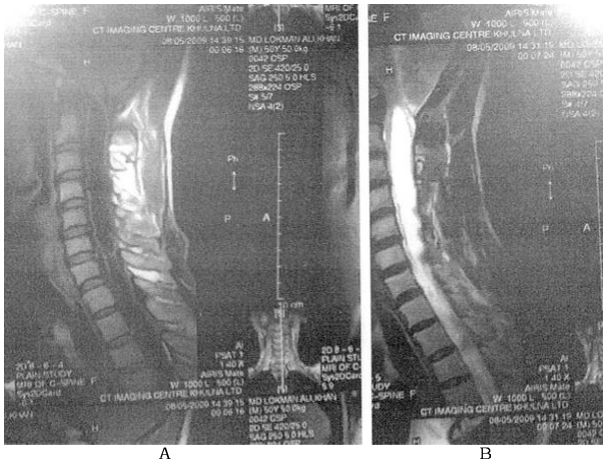
Bang Med J (Khulna) 2010; 43 : 21-23

Case Report

A 50 years old male patient was admitted in Khulna Medical College Hospital, Khulna with history of weakness of both upper limbs followed by weakness of both lower limbs. He developed wasting of muscles in his upper back region. He had also history of occasional muscle twitching in different areas of the body. All these symptoms started gradually and increased day by day without remission over a period of 1.5 years. There was no history of bladder or bowel dysfunction. There was no disturbance of sensation. He gave no history of trauma to the head or spine. There was no history of tuberculous meningitis. No history of such illness in his family.

On examination the patient was anxious. Higher psychic function was normal. There was no abnormality in speech and cranial nerves. Bulk of the muscle was reduced in all four limbs and upper back of the trunk, which was marked around the shoulder girdle. There was no fasciculation. Muscle tone was diminished in all four limbs. Muscle power was grade-III in both upper limbs and grade-IV in both lower limbs. Deep tendon reflexes were absent except the knee jerk which was normal in both sides. Plantar response was flexor in both sides. There was no ankle clonus. All modalities of sensations were intact. There were no cerebellar signs.

All the routine investigations including biochemical parameters such as CBC, PPBS, S. Creatinine, Serum T3 T4 TSH, Serum creatine phosphokinase (CPK) were normal. NCS and EMG of all limbs reveals normal findings. Radiological survey of spine was normal. MRI of cervical and dorsal spine (Fig - I) revealed a large syrinx extending from cervico-medullary junction to sixth dorsal spine with herniation of cerebellar tonsil through foramen magnum. With these findings this case was diagnosed as syringomyelia with atypical presentation.



A B

Fig 1 : MRI (A=T1 Image, B=T2 (Image) of cervical and dorsal spine showing a large syrinx extending from the cervico-medullary junction to the sixth dorsal spine.

Introduction

Syringomyelia is defined by Murry et al in1933 as the dilatation of the central canal of the spinal cord or formation of abnormal tubular cavities in the substance of the spinal cord.1 The term hydromyelia was first used by Oliver d'Angers in 1827 as a cystic dilatation of the central canal of spinal cord.2 It may be developmental as happens in chiari malformation, acquired due to intramedullary spinal cord tumour, spinal cord injury or adhesive arachnoiditis. Occational cases are idiopathic.3,4

Syringomyelia manifest classically as the triad of segmental dissociated sensory loss (loss of temperature and pain sensation and preservation of touch, joint position and vibratory sensation) in upper extremities, weakness with wasting of the small muscles of hands and pyramidal signs in the lower extremities.5, 6,7 Loss of pain sensation is the most common early neurological manifestation in syringomyelia with chiari-1 malformation.8

1. Syed Mozammel Hossain FCPS, Assistant Professor, Department of Surgery, Khulna Medical College, Khulna.
2. AKM Rabiul Iqbal DA, Assistant Professor, Department of Anaesthesiology, Khulna Medical College, Khulna.
3. Sk Farid Uddin Ahmed DA, Consultant, Department of Anaesthesiology, Khulna Medical College Hospital, Khulna.
4. Md Waliur Rahman FCPS, Resident Surgeon, Khulna Medical College Hospital, Khulna.

The case reported in this article was without any pyramidal signs and sensory loss. There was only muscle wasting and weakness of the all four limbs, This is an unusual presentation of syringomyelia.

Discussion

Syringomyelia in the cervical and dorsal spinal cord produce the clinical features of weakness and wasting of muscles of hands and arms with segmental sensory loss of dissociated type. Finally spastic paraplegia and ataxia of leg occurs due to involvement of corticospinal tract and posterior columns in the cervical region.9,10 Our case presented with flaccid quadriparesis in the absence of pyramidal signs in lower limbs and there was no sensory impairment.

On the basis of clinical features this case was thought to be a patient of motor neurone disease (MND) or peripheral neuropathy or myopathy. But Nerve Conduction Study (NCS), Electromyogram (EMG) and serum creatine phosphokinase (CPK) were normal. Radiological survey of spine was also normal. Then MRI of cervical and dorsal spine of this patient was done which revealed a large cavity, the syrinx extending from the cervicomedullary junction to sixth dorsal spine with herniation of cerebellar tonsil through foramen magnum. So the case was diagnosed as syringomyelia and this was an atypical presentation, because of the absence of pyramidal signs and sensory loss.

Atypical presentation of syringomyelia has been reported with absence of sensory loss in the presence of amyotrophy and spastic paraplegia. MRI of cervical spine of the patient revealed high cervical cord tumour (ependymoma) with secondary syrinx formation.9 Exceptionally there may be segmental dissociated sensory impairment only with preservation of motor function. Rarely cases have been recorded with spastic paraplegia only with hydrocephalus and hydromyelia.10

In the case reported there was cerebellar tonsilar herniation which is seen in chiari type-1 malformation. Acquired hind brain herniation through foramen magnum leads to syringomyelia by chronic spinal drainage of CSF or by high flow CSF shunting.11,12 In chiari type-1 malformation cerebellar tonsilar herniation occurs through the foramen magnum. A thickened band of dura mater exists at the level of the craniovertebral junction. This thickened band of dura mater causes a dissociation of CSF pressure between cranial cavity and spinal canal resulting in spinal cord cavitation. 13-15 Circulatory disturbance also occurs in the teritories of posterior spinal arteries and contributes to the formation of syrinx in spinal cord.16

In this reported patient there was no pyramidal signs in lower limbs and no sensory loss. It is a rare presentation of syringomyelia. Possibly the progression of syrinx formation was very slow which causes spliting of long tract fibres. So the sensory fibres escaped and also the pyramidal fibres remained intact.

A long term study is required to find out the exact aetiology and pathogenesis for developing such a long syrinx without pyramidal tract lesion and without sensory loss.

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