

Dissociated sensory loss and muscle wasting in a young male with headaches: syringomyelia with type 1 Arnold-Chiari malformation

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DESCRIPTION

A 21-year-old right-handed man, with no medical history of note, presented to neurosurgery outpatient department in Pakistan, with a 5-year history of numbness and pain in his upper limbs. The patient gave a troubled account of constant, current-like pain, tracking down his arms and sometimes over his chest, which frequently disturbed his sleep. The numbness he experienced made it difficult to turn pages of the newspaper. Upon further questioning, he also admitted to suffering with occipital headaches for a number of years. Examination of the upper limbs revealed wasting of the thenar and hypothenar eminences of the left hand and loss of pain and temperature sensation in the C3-T2 dermatomes, bilaterally. The remainder of the neurological examination was normal. An MRI spine was performed, revealing a long, fluid-filled central

cavitation, with accompanying descent of the cerebellar tonsils (figure 1). Syringomyelia with type 1 Arnold-Chiari malformation was diagnosed and the patient was offered decompressive surgery.

Syringomyelia results from the development of a syrinx within the spinal cord, most commonly in the cervical segment. It is frequently associated with a type 1 Chiari malformation of the spine, in which the cerebellar tonsils descend below the foramen magnum. The exact length of descent for diagnosis is controversial, as up to a third of normal participants may harbour this finding,¹ though >5 mm is considered pathological.² In the presence of a Chiari-malformation, first-line management is usually posterior fossa decompression, comprising a C1 laminectomy and partial occipital craniectomy, with or without opening of the dura.

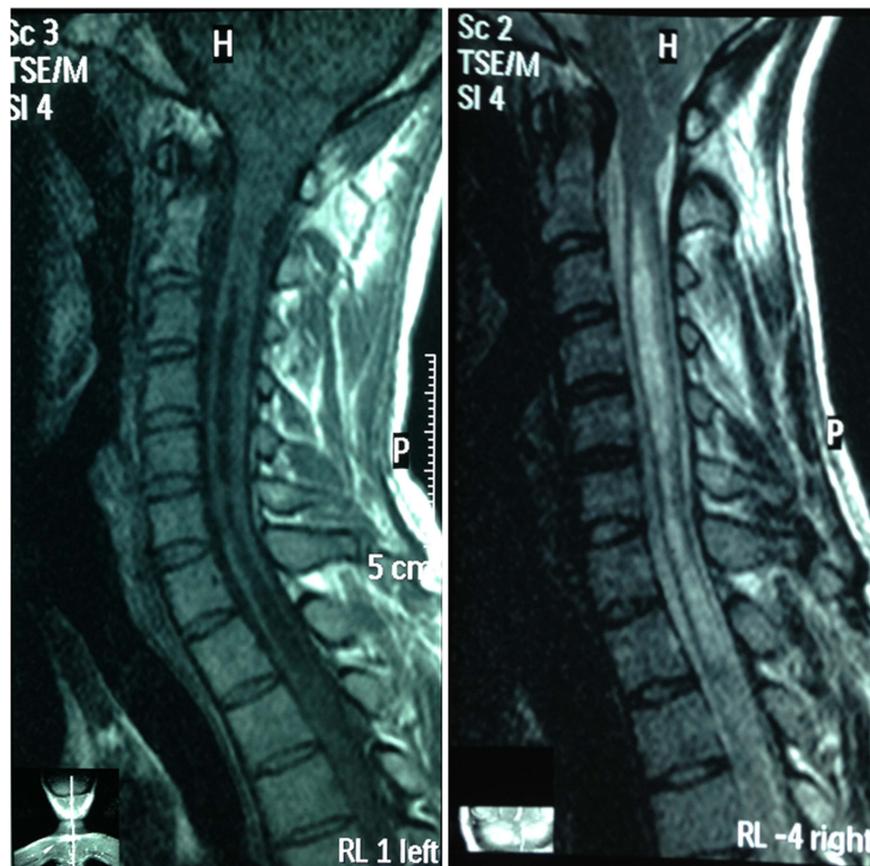


Figure 1 MRI spine showing a fluid-filled, central cavity. The presence of fluid can be deduced the long hyperintensity on T2-weighted imaging (right). Descent of cerebellar tonsils below the level of the foramen magnum is also apparent.



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Learning points

- ▶ Dissociated sensory loss in a young male should lead to suspicion of syringomyelia.
- ▶ Syringomyelia is frequently associated with Chiari malformations of the spine, indicating imaging of the cervico-medullary junction in all patients.
- ▶ The length of cerebellar tonsillar herniation needed for a diagnosis of Chiari-1 malformation varies, but 5 mm is considered pathological.

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