

Extensive Hydromyelia

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Hydromyelia is frequently associated with dissociated sensory loss, scoliosis, and upper limb weakness. This report describes the case of a 9-year old male with an extensive hydromyelia associated with a spinal cord tumor and an oligosymptomatic clinical presentation.
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Introduction

Hydromyelia is a dilatation of the spinal central canal that communicates with the intracranial cerebrospinal fluid compartment [1]. It is usually associated with developmental abnormalities at the foramen magnum and posterior fossa, spinal arachnoiditis, and spinal cord tumor [2]. This report presents a case of an extensive hydromyelia associated with a spinal cord tumor and an oligosymptomatic clinical presentation.

Case Report

A previously healthy 9-year-old male was admitted because of back pain at the T₅ level. Six months before admission, he developed a dull pain in the dorsal region at the T₅ level, which was nonradiating, with no aggravating or alleviating factors and whose intensity gradually increased over time. One month before admission, he also developed an orthostatic headache and a mild gait imbal-

ance. He denied any bowel or urinary symptoms. The neurologic examination revealed no cranial nerve deficits, but a slight horizontal nystagmus at extremes of gaze was observed. There was a mild symmetric tetraparesia with decreased deep tendon reflexes and flexor plantar responses. Sensory examination disclosed no abnormalities. Although there was no dysmetria, an ataxic gait was observed.

A magnetic resonance imaging study of the spine revealed an irregular lesion at T₃-T₄ level and an extensive dilatation of the central canal from the fourth ventricle to the filum terminale suggestive of hydromyelia (Fig 1). A Chiari malformation was not present; brain magnetic resonance imaging disclosed no abnormalities except for the fourth ventricle dilatation with posterior displacement of the cerebellar vermis. A biopsy was performed at T₄ level, and the histologic examination of a dorsal surface fragment was suggestive of a pilocytic astrocytoma. Partial resection of the lesion was performed and chemotherapy planned, but the patient was transferred to another institution and was lost to follow-up.

Discussion

Hydromyelia is associated with an insidious onset and a slow progression of the symptoms. Dissociated sensory loss is a common feature and, in pediatric patients, upper limb weakness and scoliosis are frequently observed [3,4]. In the present case, despite the dilatation of the central canal observed in all the spinal cord extension, the patient presented with mild motor symptoms and no sensory deficits. The gait ataxia could be explained by a compression of the cerebellar vermis and the hypoactive deep tendon reflexes by the involvement of the anterior horn cells.

There is no consensus about the pathogenesis of hydromyelia. A developmental defect in the medullo-cervical junction that prevents the free flow of the cerebrospinal fluid is a proposed explanation. The normal cerebrospinal fluid pulse wave generated by the arterial systolic pulsation at the choroid plexus would be directed down the central canal leading to progressive dilatation. Alternatively, hydromyelia would be an acquired anomaly in which dissociation between cranial and spinal cerebrospinal fluid pressures (spinal pressure

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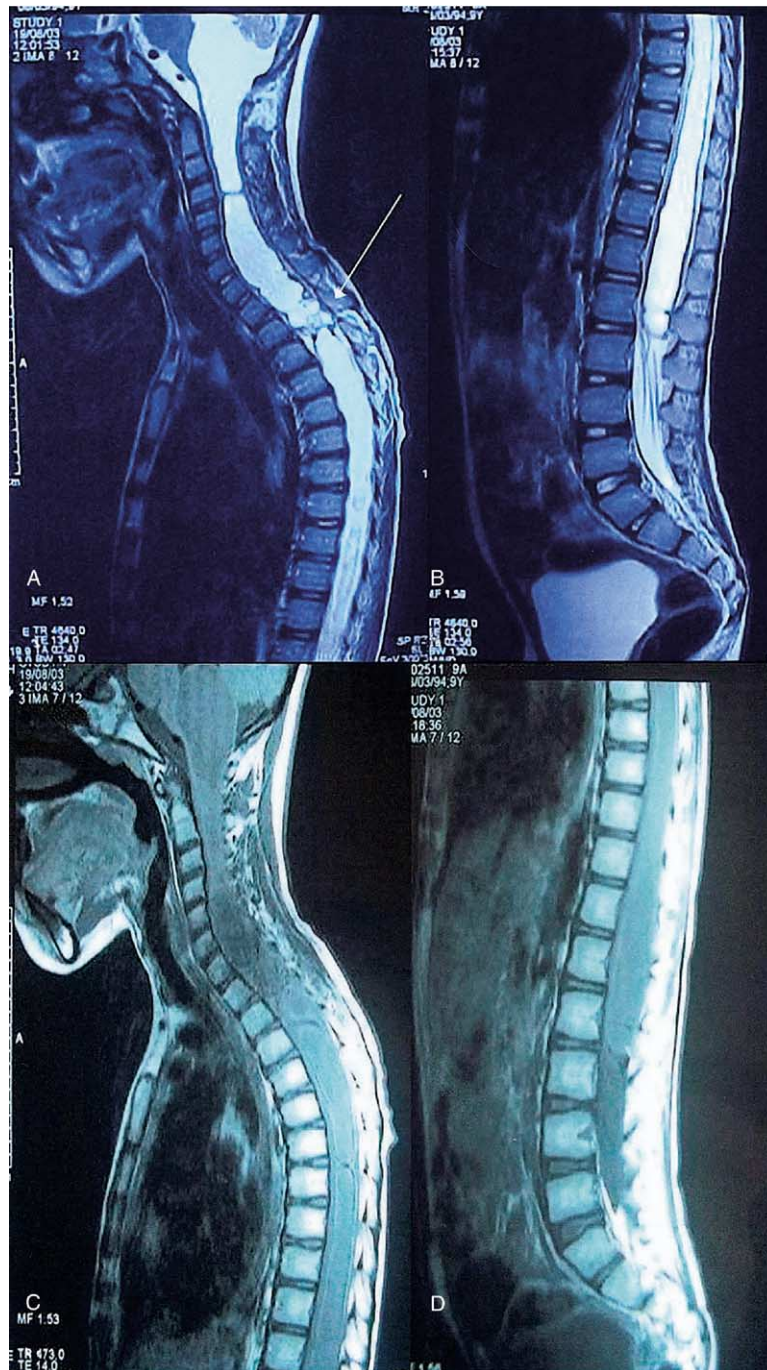


Figure 1. T_2 -weighted (panels A and B, TR/TE= 4640/134 ms) and T_1 -weighted (panels C and D, TR/TE= 473/14 ms) images of the cervical and thoracic and lumbar segments reveal an extensive dilatation of the central canal of the spinal cord from the fourth ventricle to the filum terminale and an irregular intramedullary lesion at T_3 level (white arrow). An enlargement of the foramen magnum is also observed (panels A and C).

negative to cranial pressure), produced by an obstruction anywhere in the spinal axis, draws fluid into the cord, resulting in central canal dilatation [3]. Recently, however, it has been proposed that hydromyelia associated with Chiari II malformation would be secondary to a molecular genetic defect rather than a mechanical problem [5].

We speculated that the clinico-radiologic mismatch observed in the present case can be explained by the slow growing nature of the tumor leading to a subtle accumulation of fluid and gradual displacement of the neural structures, as well as the plasticity of the developing nervous system which adjusts to such deviations of the normal anatomy.

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References

- [1] **Johnston I**, Teo C. Disorders of CSF hydrodynamics. *Childs Nerv Syst* 2000;16:776-99.
- [2] **Wisoff JH**. Hydromyelia: A critical review. *Childs Nerv Syst* 1988;4:1-8.
- [3] **Gower DJ**, Pollay M, Leech R. Pediatric syringomyelia. *J Child Neurol* 1994;9:14-21.
- [4] **Iqbal JB**, Bradey N, Macfaul R, Cameron MM. Syringomyelia in children: Six case reports and review of the literature. *Br J Neurosurg* 1992;6:13-20.
- [5] **Sarnat HB**. Regional ependymal upregulation of vimentin in Chiari II malformation, aqueductal stenosis, and hydromyelia. *Pediatr Dev Pathol* 2004;7:48-60.