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Case Report

A patient with an arachnoid cyst with secondary syringomyelia presenting with pure motor symptoms

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ABSTRACT

Syringomyelia is a condition in which a fluid-filled cavity is formed within the spinal cord; it can be asymptomatic and when symptomatic it most often presents with sensory disturbances. In rare cases, a syrinx develops secondary to an arachnoid cyst. We present a patient suffering from pure motor symptoms due to an arachnoid cyst with secondary syringomyelia. After surgery, an improvement of symptoms was seen.

KEYWORDS: arachnoid cyst, syringomyelia, surgery, unilateral motor symptoms

CASE REPORT

A 63-year old male presented with one year lasting progressive loss of strength in his right hand with muscle wasting and difficulty in writing. He experienced no pain, sensory loss nor paresthesia. He had no complaints of the lower extremities and there were no problems with micturition or defecation.

Neurologic examination showed a substantial atrophy of the m. adductor pollicis and paresis of the ab- and adductors of the fingers (Medical Research Counsil (MRC) grade 4) without sensory deficit. Examination of the lower extremities was normal. Magenetic Resonance Imaging (MRI) of the brain showed no abnormalities. MRI of the cervical spine documented a syrinx from C6 to Th3, as well as a lesion dorsally of the spinal cord at Th3-Th4 with compression on the myelum, suspected of an arachnoid cyst (Fig. 1A and B). Because of the absence of sensory deficit, an electromyogram (EMG) was performed which showed no signs of anterior motor horn disfunction, nor peripheral nerve abnormalities.

The progressive weakness of the patient's right hand was most likely caused by the syrinx and arachnoid cyst, and a neurosurgeon (MA) was consulted for surgical treatment. A laminectomy Th2-Th4 was performed, and a dorsally located arachnoid cyst at Th3-Th4 was excised and fenestrated. In addition, a myelotomy at the level of Th2-Th3 was performed to release cerebrospinal fluid into the arachnoid space; no drain was placed in the syrinx.

Following surgery, motor functioning of the right hand recovered completely, and improvement of atrophy was seen. Postoperative MRI at 6 months showed a decrease of syringomyelia and no signs of recurrence of the arachnoid cyst (Fig. 2A and B). The patient was discharged from further follow-up.

DISCUSSION

Syringomyelia is defined by the presence of a fluid-filled cavity within the spinal cord, which is usually found at a cervical or thoracic level [1]. It can be asymptomatic although patients usually present with pain and sensory disturbances in the upper extremities caused by damage to the centrally crossing fibers of the spinothalamic tract [2]. Motor deficit of the extremities may also be

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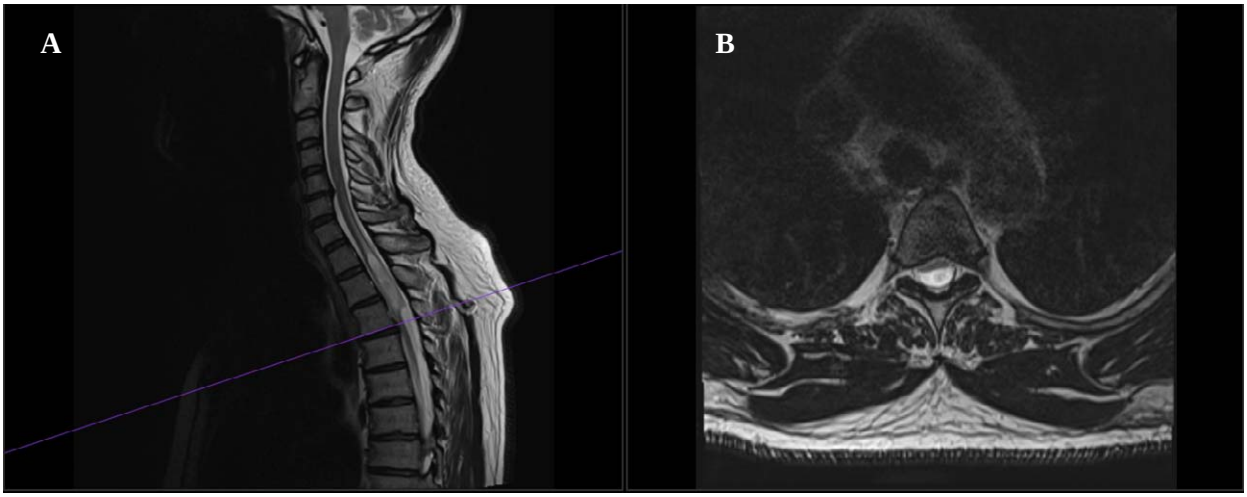

Fig. 1A and B. (pre-operative images): on the sagittal view a syrinx extending from C6 to Th3 can be seen, as well as an arachnoid cyst extending caudally from Th3. An axial view at the level of Th3-Th4 shows a dorsal arachnoid cyst.

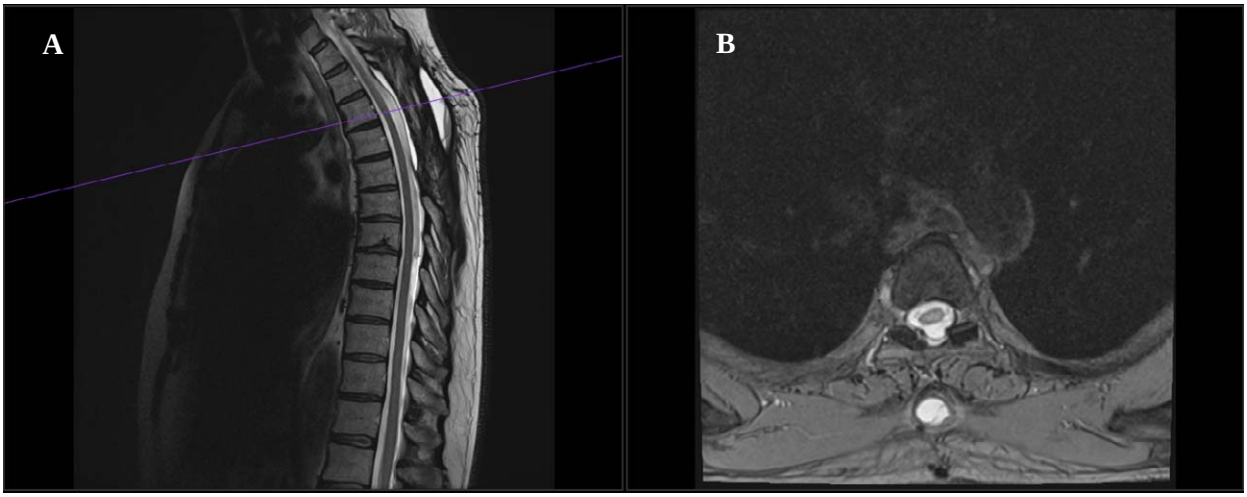

Fig. 2A and B. (post-operative images): six months post-operatively no arachnoid cyst is present and there is a substantial decrease of the syringomyelia.

present. Syringomyelia can be caused by several entities like Arnold Chiari malformation, hydrocephalus, trauma, infection and inflammation. The pathogenesis of syringomyelia has yet to be fully elucidated. It is hypothesised that disturbance of normal anatomy may lead to an aberrant flow of the cerebrospinal fluid which may cause a change in distribution of extracellular fluid, eventually leading to the formation of a syrinx. Treatment consists of surgical intervention focussing on the cause of syringomyelia.

Arachnoid cysts are seen less frequently than syringomyelia. They can be either asymptomatic or lead to complaints such as back pain, chest discomfort, numbness of the lower extremities, gait disturbance and urinary incontinence [3]. The combination of syringomyelia and arachnoid cysts is even more uncommon [4, 5]. The exact incidence

of syringomyelia in patients with arachnoid cysts is unknown. It has a predominance in males (78.1%), who most often present with walking difficulty and urinary and fecal incontinence. Most cysts are located at the thoracic level (84.4%), and in most cases the syrinx is located at the same level of the arachnoid cyst, or one level below. In most reported cases, the surgical treatment consisted of solely excision or fenestration of the arachnoid cyst, although in 9 patients the syrinx was also treated by fenestration or placement of a shunt. Most cases (89.7%) showed radiological improvement after surgery as well as neurological improvement (87.5%).

In this case report, we presented a patient with pure motor symptoms due to a cervicothoracic syrinx secondary to a dorsally located arachnoid cyst. Our case presentation is unusual because patients with a syringomyelia usually present with pain and other sensory symptoms, while literature shows that most patients with a combination of syringomyelia and an arachnoid cyst most often present with gait difficulty and incontinence. The basic surgical treatment of an arachnoid cyste with syringomyelia involves removal of the cyst which may restore normal cerebrospinal fluid flow leading to reduction of syringomyelia. In our case, we also treated the syrinx by fenestration. There is no consensus on the best surgical treatment, although the combination of cyst removal and fenestration of the syrinx has resulted in good neurological and radiological outcome in the presented case.

AUTHOR CONTRIBUTION

M. A. and J. S. have written the manuscript and approved the final version.

CONFLICT OF INTEREST STATEMENT

The authors have no disclosures deemed to be relevant to the submitted manuscript.

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