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Multiple sclerosis and syringomyelia – a case report

Stwardnienie rozsiane i syringomielia – opis przypadku

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Abstract

Syringomyelia is associated with more than two-thirds of Chiari malformation type I cases, and rarely with intramedullary neoplasm or post-traumatic cavitations. Some authors assume that syringomyelia, sometimes observed in patients with multiple sclerosis, is more likely to be a consequence of inflammatory-demyelinating pathology of the spinal cord. We describe a case of a 23-year-old patient who was diagnosed with multiple sclerosis according to the McDonald criteria with syringomyelia and Chiari malformation type I. It to be a very rare case of co-occurrence of syringomyelia and multiple sclerosis as two separate unrelated independent diseases.

Keywords: multiple sclerosis, syringomyelia, Chiari malformation

Streszczenie

Jamistość rdzenia polega na nieprawidłowym poszerzeniu kanału centralnego rdzenia kręgowego i jest związana w ponad 2/3 przypadków z zespołem Arnolda–Chiariego typu I, rzadziej z guzem wewnątrzrdzeniowym lub przebytym urazem – jako jego powikłanie. Według niektórych autorów rzadko występujące jamy syryngomieliczne obserwowane u pacjentów ze stwardnieniem rozsianym są następstwem zmian demielinizacyjnych w rdzeniu kręgowym. W pracy przedstawiamy przypadek 23-letniego pacjenta z rozpoznanym zgodnie z kryteriami McDonalda stwardnieniem rozsianym oraz jamistością rdzenia kręgowego i zespołem Arnolda–Chiariego typu I. Opisywany przypadek potwierdza, iż w rzadkich przypadkach stwardnienie rozsiane może współwystępować z jamistością rdzenia kręgowego jako dwie oddzielne, niezwiązane ze sobą jednostki chorobowe.

Słowa kluczowe: stwardnienie rozsiane, jamistość rdzenia, zespół Arnolda–Chiariego typu I

INTRODUCTION

Syringomyelia is an abnormal enlargement of the central canal of the spinal cord and is associated with more than two-thirds of Chiari malformation type I cases, rarely with intramedullary neoplasm or post-traumatic cavitations and sometimes needs surgical intervention. Some authors have assumed that syringomyelia, sometimes observed in patients with multiple sclerosis (MS), is more likely to be a consequence of inflammatory-demyelinating pathology of the spinal cord, rather than co-occurrence of two different diseases (Weier et al., 2013, 2008).
To our best knowledge, the coexistence of MS and syringomyelia with Chiari malformation type I in the same patient has never been reported and this is the first report which describes such a patient.

CASE REPORT

The first symptom in a 23-year-old man, which occurred in December 2012, was muscle weakness of the right lower limb. This symptom resolved without treatment. However, in September 2013, the following occurred: pyramidal weakness of the lower limbs, loss of balance and sensory disturbances over the trunk and the lower limb on the left side. In addition, the patient had had a history of back pain for several years (thoracic region of the spinal cord), a knee bone defect (congenital dislocation of patella) and scoliosis at the Th–L segment of the vertebral column. On neurological examination, the patient presented with pyramidal weakness as tetraparesis which affected the lower limbs more severely, bilateral cerebellar signs and loss of light touch sensation over the left side below Th8 level. Clinical examination did not show typical signs of syringomyelia-dissociated sensory loss. Magnetic resonance imaging (MRI) of the brain revealed periventricular and subcortical white matter lesions in both hemispheres, the corpus callosum, the cerebellar peduncles, the brainstem- medulla, multiple hyperintense foci on T2 weighted and fluid-attenuated inversion recovery (FLAIR) images, without contrast enhancement. In addition, the cerebellar tonsils descended 8 mm below the foramen magnum (Fig. 1). Spinal cord MRI revealed hyperintense band on T2-weighted images corresponding to syrinx formation from C5 to Th12 and

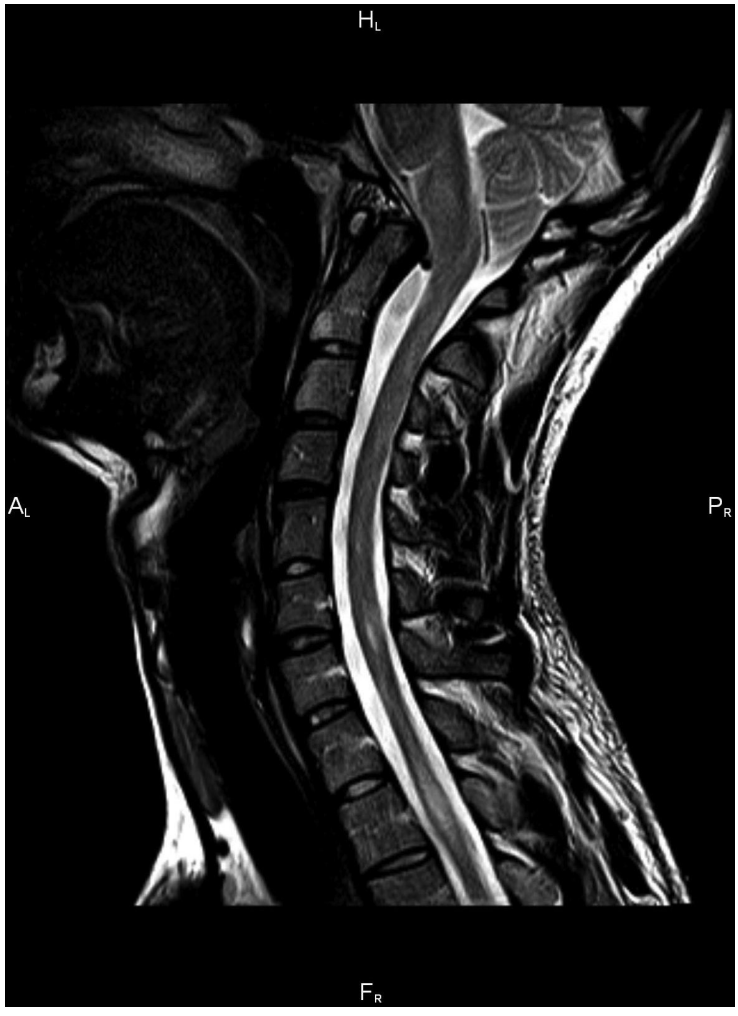

Fig. 1. MRI of the cervical spinal cord, sagittal plane, T2-weighted images. Cerebellar tonsils descended below the foramen magnum

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MS cord lesion on the left side at Th2 level (Fig. 2). On sagittal images the cord appeared slightly distended and the cavity size was 3–4 × 6 mm (APxPR) in the widest place at the level C6–C7. Cerebrospinal fluid (CSF) examination revealed 1 cell, a protein level of 106 mg/dL, and oligoclonal bands. Antibody testing in serum and in CSF ruled out Lyme disease. MRI of the lumbar spine did not reveal any abnormality. Treatment with a steroid resulted in improvement – decreased pyramidal weakness, sensory deficit and less extent cerebellar symptoms. After neurosurgical consultation, CSF flow imaging with a phase-contrast MR technique was performed. Quantitative CSF flow velocity in the aqueduct was 2–3 cm/s (normal value: 5–8 cm/s). The CSF flow through the fourth ventricle towards the foramen magnum into the cerebellomedullary cistern was normal. As in MRI, the lowering of the tonsils was moderate and there were no signs of compression on the lower brain stem and cervical spine, therefore the patient was not qualified for a neurosurgical procedure.

Because neurological symptoms were steroid responsive, MS was diagnosed, and we started treatment with a disease-modifying therapy (interferon beta). In the view of the above, the patient was diagnosed with MS, syringomyelia, and Chiari malformation type I.

The neurological condition of the patient was stable to 2015 (Expanded Disability Status Scale, EDSS 4.0); in December a relapse occurred with a good response to the steroid treatment. In March 2017, another relapse occurred, and we switched interferon beta to dimethyl fumarate. Repeated brain and spinal cord MRI did not reveal new MS lesions and syrinx progression.

DISCUSSION

Syringomyelia is a rare disease with a prevalence of 8.4/100,000 to 0.9/10,000 in the normal population (Ferrero Arias and Pilo Martín, 1991). In patients with MS, syringomyelia is described in case reports or small case series as incidental findings of spinal cord pathology (Basedow-Rajwich et al., 1995; Ferrero Arias and Pilo Martín, 1991; Larner et al., 2002; Weier et al., 2013, 2008). Weier et al. (2008) reported on nine patients with MS and syringomyelia, who participated in a study of 202 (4.5%) patients with MS. The next paper by these authors was associated

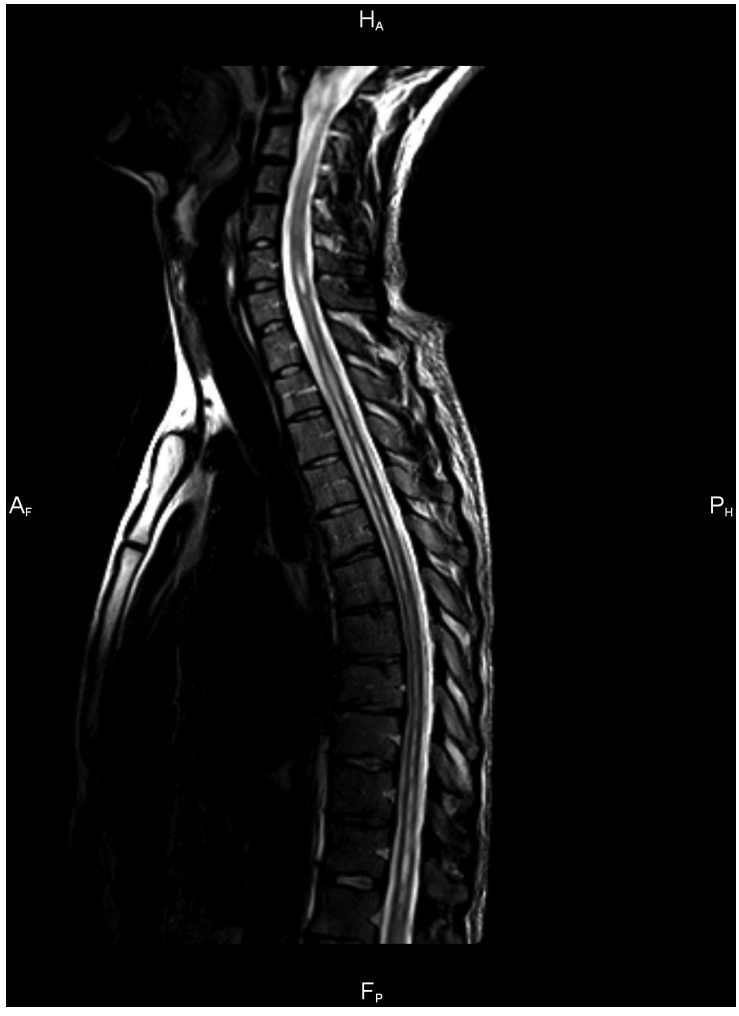

Fig. 2. MRI of the cervical and thoracic spinal cord, sagittal plane, T2-weighted. Syrinx formation at the cervical and thoracic spinal cord

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with clinical and MRI characteristics of syringomyelia in a six-years follow-up observation of patients with MS and syringomyelia (Weier et al., 2013). Syringomyelia was not associated with a history of trauma or spinal cord surgery. Clinical examination showed none of the typical symptoms associated with syringomyelia. None of the patients had abnormalities of the craniocervical junction. The syrinx formations were located only in the thoracic or lumbar cord. The extension of each cavity ranged from less than one vertebral body up to more than five vertebral bodies (2.5–17 cm length). Only one patient had 17 cm syrinx length, other 15 cm and 9 cm, and the rest of patients – less than 5 cm. The authors assumed that syringomyelia is not a coincidental finding in patients with MS but is associated with the spinal cord inflammatory, which is a demyelinating pathology. The six-year follow-up revealed stable findings regarding the size and shape of the syrinx. According to the authors, syringomyelia cannot be seen as a poor prognostic sign in MS (Weier et al., 2013, 2008).

In our patient, syrinx formation ranged from C5 to Th12 level, which is typical for syringomyelia, and cerebellar tonsils descended below the foramen magnum. Examination of the patient did not reveal dissociated sensory loss, although clinical features of syringomyelia were found i.e. typical pain syndrome and bone defects (congenital dislocation of patella and scoliosis). It seems that the syrinx in our patient was too widespread as for the one resulting from an inflammatory-demyelinating pathology. In the cases described by Weier et al. (2013, 2008), there were no peculiar clinical histories or uncommon clinical findings suggesting other diagnosis or other pathologies. Therefore, authors assumed that syringomyelia was not a coincidental finding but was related to spinal cord involvement in MS (Weier et al., 2013, 2008). In our patients, demyelination process did not exacerbate syringomyelia and syrinx showed no tendency to change in size or shape over 4 years. Additional symptoms or diseases like bone defects, Chiari malformation type I, and extensive syrinx including cervical and thoracic part of the spinal cord might be considered separate diseases.

We would like to emphasise that it is worthy to look for an alternative pathology when a patient with MS develops a new neurological deficit, because some of them may need a different treatment than MS treatment.

Conflict of interest

The authors do not report any financial or personal affiliations to persons or organisations that could adversely affect the content of or claim to have rights to this publication.

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