

Pathophysiology of presyrinx state associated with Chiari malformation depicted on magnetic resonance imaging: cerebrospinal fluid infiltration pathways and interstitial edema

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A 33-year-old man presented with nuchal pain, dysesthesia in the fingers, and hyperreflexia. A 1.5-T magnetic resonance imaging (MRI) revealed Chiari malformation and spinal cord edema dorsal to a ventrally-located thin syrinx. On axial views, the edema was depicted as a wedge-shaped signal between the dorsal aspect of the spinal cord and the syrinx with T1 hypointense and multiple linear T2 hyperintense signals. Diffusion-weighted imaging (DWI) showed no hyperintense signal in the edema. Foramen magnum decompression improved his symptoms. Although the cord edema improved somewhat, a syrinx partly developed.

The linear T2-hyperintense signals were compatible with the dorsal root and Virchow-Robin's spaces between the subarachnoid space and central canal, the putative cerebrospinal fluid infiltration pathways forming the syrinx. The finding on DWI proved that the edema was interstitial. An accumulation of higher resonance imaging scans of the presyrinx state at various stages is needed to elucidate the *in vivo* syrinx formation process.

Key words: Chiari malformation, magnetic resonance imaging, pathophysiology, presyrinx state, syringomyelia

Introduction

The presyrinx state, an entity established after the introduction of magnetic resonance imaging (MRI), was first described by Fischbein et al. in 1999.¹ It is associated with various pathologies resulting in disturbance of the cerebrospinal fluid (CSF) flow such as Chiari malformation, basilar arachnoid adhesions, hydrocephalus, meningitis, cervical spondylostenosis, and basilar impression.¹ On MRI scans, the presyrinx state is depicted as enlargement of the spinal-, primarily the cervical cord, with T1 and T2 prolongation (the T1 signal is not as low as the CSF signal and manifests a dull margin) but no frank cavitation.¹ These are findings thought to precede the formation of true syringomyelia. Although there is an association with nonspecific myelopathic symptoms, they are usually improved by surgical release of the CSF obstruction.¹ Progression to syrinx formation after surgical release of the CSF obstruction is rare.¹

We present a case of presyrinx associated with Chiari malformation, in which MRI findings supported a pathophysiological hypothesis of syrinx development.

Case study

A 33-year-old man had a 6-month history of nuchal and occipital pain that was exacerbated by coughing, and a 3-month history of dysesthesia in the left fingers.

At presentation, neurological examination revealed markedly increased tonus and hyperreflexia in all extremities. His gait was almost normal and pathological reflexes were negative. His response to tactile and pinprick stimulation was reduced in the left hand. A 1.5-T MRI scan yielded findings of Chiari malformation and associated abnormalities in the cervical spinal cord below the foramen magnum. We noted a thin syrinx at the ventral aspect, intramedullary edema dorsal to the syrinx, and spinal cord enlargement (Figures 1A, B). On axial views, the edema was depicted as a wedge-shaped

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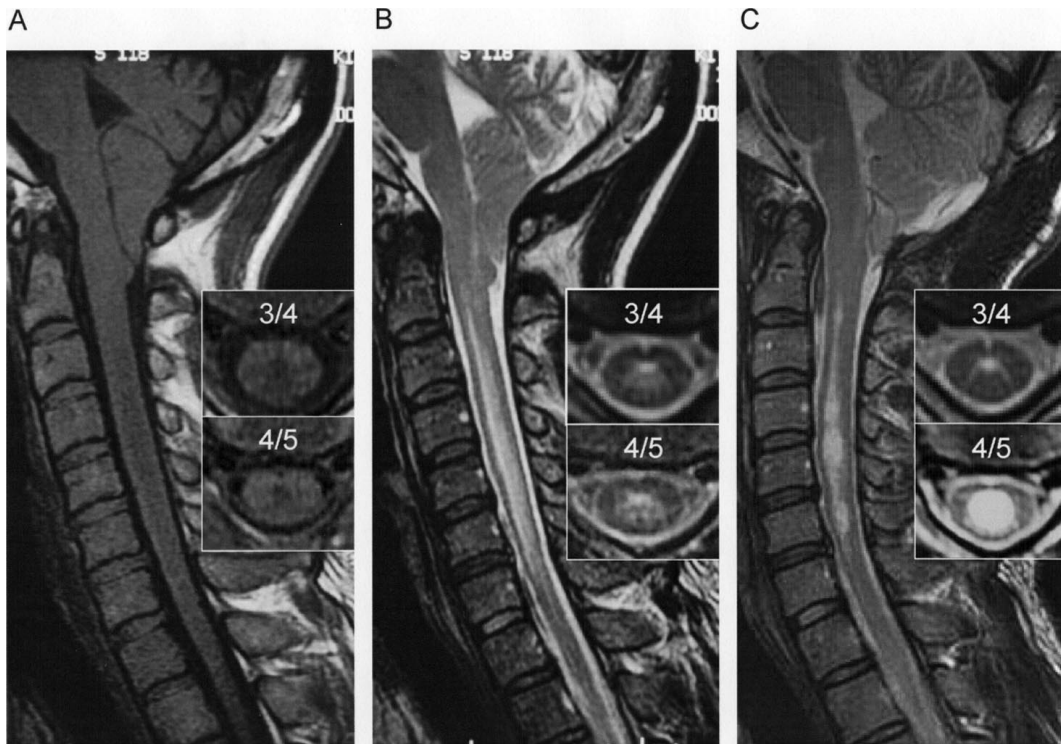


Figure 1. Sagittal T1- (A) and T2-weighted (B) MRI performed at presentation shows the Chiari malformation with tonsillar herniation. There is enlargement of the spinal cord with T1 and T2 prolongation at the C2-Th1 levels. The T1-signal is not as low as the CSF signal and manifests a dull margin. On axial views, edema is depicted as a wedge-shaped abnormal signal between the dorsal aspect of the cord and the syrinx; note the T1 hypointense- and multiple linear T2 hyperintense signals (insets). Sagittal T2-weighted MRI obtained 9 months after surgery (C). The lower tip of the tonsil has ascended and the dorsal hump has disappeared. The T2 hyperintense signals at C3/4 have disappeared, and a syrinx has developed at C4/5 (insets).



Figure 2. Sagittal DWI performed at presentation (A) and 9 months after surgery (B). There is no hyperintense signal in the region corresponding to the edema, syrinx, or cord parenchyma adjacent to the syrinx.

abnormal signal between the dorsal aspect of the cord and the syrinx. There were T1 hypointense and multiple linear T2 hyperintense signals (insets in Figures 1A, B). Diffusion-weighted imaging (DWI) showed no hyperintense signal in the region corresponding to the edema (Figure 2A).

We performed foramen magnum decompression surgery with duraplasty and removal of the posterior arch of the atlas. The arachnoid was partly torn, but it was covered with a dural substitute, an expanded polytetrafluoroethylene sheet.

At 9 months after the operation, all his symptoms except the dysesthesia in the left fingers were resolved. MRI showed improvement of the cord edema in the rostral part, but a syrinx had developed in the caudal part (Figure 1C). On DWI there was no hyperintense signal in either the syrinx or adjacent cord parenchyma (Figure 2B). Subsequent follow-up was not possible because the patient moved.

Discussion

Various pathophysiologies have been proposed for syrinx formation. Syringomyelia was first thought to be attributable to the forceful diversion of CSF from the fourth ventricle into the central canal of the cord due to obstruction of the outlets of the fourth ventricle² or the development of a pressure gradient between the intracranial and intraspinal spaces.³

More recently, the hypothesis first posed by Ball and Dayan⁴ that the CSF in the subarachnoid space enters the central canal through the dorsal roots or Virchow-Robin's spaces, has been accepted because continuity between the fourth ventricle and syrinx is rarely observed on MRI scans. Based on the communication with the central canal, syringomyelia can be classified into three pathologic types,⁵ i.e., the central canal syringes that communicate with the fourth ventricle in association with hydrocephalus; the noncommunicating central canal syringes associated with congenital and acquired disorders including Chiari malformations, arachnoiditis, and extramedullary compressive lesions; and the extracanalicular (parenchymal) syringes that are associated with trauma, infarction, and hemorrhage.

The spinal subarachnoid space is continuous with the central canal through a series of interconnecting, perivascular and interstitial spaces with a minimum diameter of 150-200 μ m.^{5,6} Normally, CSF flows from the subarachnoid- to the subpial- before entering the perivascular space. It circulates through the cord parenchyma toward the central canal, but may also flow

in reverse, as these forces are relatively balanced.¹ In the presence of the Chiari malformation, the tonsil moves downward rapidly during systole, and the increased pressure drives the CSF into the cord parenchyma to form a syrinx, and the extent of the syrinx formation by this mechanism may be related to different degrees of central canal stenosis.¹ The underlying pathology of central canal stenosis involves the proliferation of subependymal gliovascular nodules. The incidences of which increase with age.⁷ In the presence of central canal stenosis, CSF driven into the cord parenchyma by increased CSF pressure cannot accumulate in the central canal, and it diffuses through the cord parenchyma, resulting in cord enlargement and edema, i.e., the presyrinx state.¹

The wedge-shaped abnormal signal we observed between the dorsal and ventral aspect of the cord on axial views, depicted as T1 hypointense and linear T2 hyperintense signals, was compatible with CSF infiltration into the dorsal root and the Virchow-Robin's spaces between the subarachnoid space and the central canal, and the putative CSF infiltration pathways resulting in syrinx formation.^{5,6} The lack of a hyperintense signal on DWI in the area corresponding to the edema and beside the developing syrinx, confirms that the pathology of the reversible edema was interstitial rather than cellular with restriction of water diffusion within the cells and cell swelling due to an influx of extracellular water and Na⁺.⁸ Spinal cord infarction is a representative pathology of cellular edema.⁹ The present interstitial edema disappeared at C3/4 while it progressed to syrinx formation at C4/5 after decompression surgery. This regional progression to syrinx formation might contribute to residual disturbance of the CSF circulation, which the surgery failed to solve. Such an unfavorable result of the surgery was occasionally observed, and in the such cases pre-existent- or surgery-induced arachnoid adhesion at the foramen magnum was considered to be the cause.¹⁰

In conclusion, we first present MRI evidence of the CSF infiltration pathways into the spinal cord resulting in syrinx formation and of the interstitial nature of the edema. The accumulation of higher resonance imaging scans of various presyrinx stages is needed for an understanding of the process underlying the formation of syringes in vivo.

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