



Case Report

Imaging evolution from “presyrinx” to syrinx in patient with spinal lipoma

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ABSTRACT

Background: The evolution of syrinx formation has rarely been documented. Here, we report a patient whose “presyrinx” evolved on successive magnetic resonance (MR) images to a mature syrinx.

Case Description: A patient had a lipoma and tethered cord at birth. At 3 weeks of age, he had undergone a partial removal of the lipoma and untethering of the spinal cord. At age 6, the thoracic MR images showed edema within the gray matter of the cord at the T7 level, consistent with a “presyrinx.” In addition, subsequent MR studies (i.e., at age 7) showed a small cavity in the right posterior horn of the cord accompanied by further expansion throughout the right-sided gray matter. Despite repeated cord untethering at age 7, the T7 parenchymal cord change evolved into a mature syrinx by age 10.

Conclusion: An infant with a lipoma/tethered cord, despite two instances of cord detethering (i.e., ages 3 weeks and 7 years), showed continued MR evolution of a “presyrinx” to a mature syrinx by age 10.

Keywords: Presyrinx, Spinal lipoma, Syrinx

INTRODUCTION

A syrinx is a fluid-filled cystic cavitation in the spinal cord parenchyma that develops due to the disturbed resorption of the extracellular fluid (ECF) from the spinal cord into the venous system.^[5,9] Early edematous cord changes, called “presyrinx,” precede mature syrinx formation.^[2,6] Here, a child with a lipoma/tethered cord who had two surgical detethering procedures (i.e., at age 3 weeks and 7 years) demonstrated magnetic resonance (MR) evolution from a “presyrinx” into a mature syrinx by age 10.

CASE REPORT

An infant presented with lipomyelomeningocele/tethered cord in the lumbosacral area at birth [Figures 1a-c]. Three weeks later, the lipoma was partially removed, and the cord was untethered [Figures 1d and e]. At age 6, MR images revealed edema in the gray matter of the spinal cord extending from T7 to T9 [Figures 2a and b]. By age 6.5, he exhibited intermittent

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lower extremity pain and weakness. The repeat MR images at age 7.25 showed partial cord cavitation which, within 9 months, expanded through the right-sided gray matter;

notably, the “lesion” did not enhance [Figures 2c-f]. Despite a repeated untethering procedure, by age 10, the “lesion” evolved into a mature syrinx [Figures 2g and h].

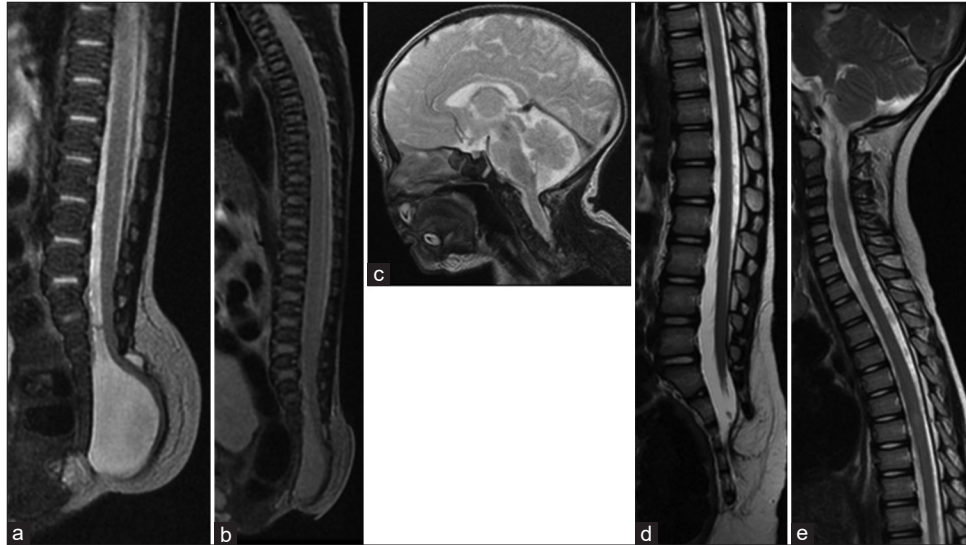


Figure 1: (a) Sagittal T2-weighted magnetic resonance (MR) image at birth showed the sacral lipomyelomeningocele. The spinal cord prolapsed from the sacral epidural space and tethered at the subcutaneous fat. (b and c) No obvious syringomyelia, Chiari malformation, and hydrocephalus were observed at birth. (d and e) Sagittal T2-weighted MR image at age 5 years showed no syrinx formation. Although the low-lying conus could be observed, the patient was asymptomatic; therefore, close observation was continued.

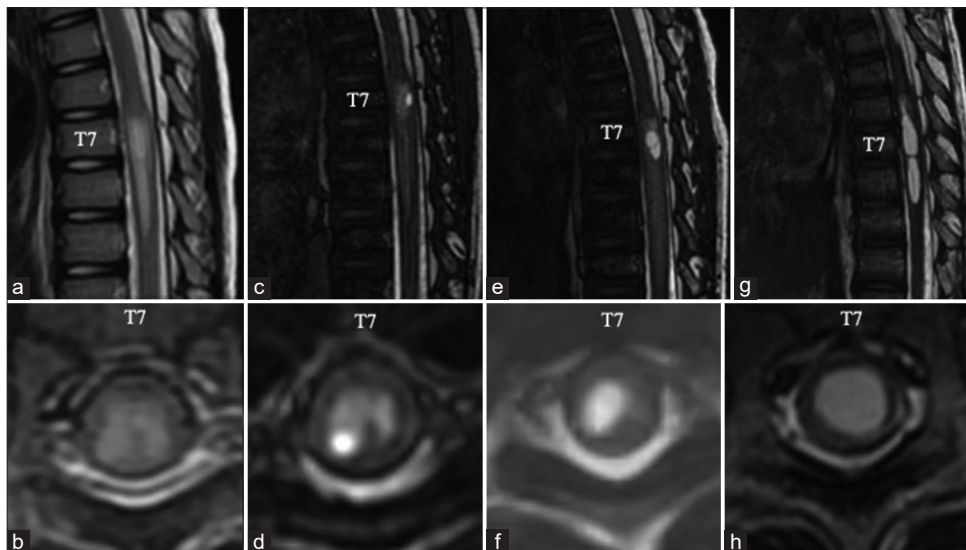


Figure 2: (a) Sagittal T2-weighted magnetic resonance (MR) image at age 5 years showing the newly emerging T2 prolongation at T7-T9. (b) Axial T2-weighted MR image showing the abnormal T2 prolongation confined to the gray matter with no frank cavitation. At this time, the patient was still asymptomatic. (c and d) Six months later, the patient complained of intermittent pain and weakness in his lower limbs. MR image showing syringomyelia cavitation in the right posterior horn at T7. (e and f) Magnetic resonance (MR) image at age 6 years showing that the syrinx was enlarged and expanded throughout the right gray matter. Since retethering was considered the cause of the intermittent lower limb symptoms and the progression of syringomyelia, a revision untethering surgery was performed. After the operation, symptoms were resolved. (g and h) However, on MR imaging 6 months after surgery, the syrinx had enlarged into the entire spinal parenchyma.

DISCUSSION

Presyrinx state

“Presyrinxes” develop due to various disorders in the cerebrospinal fluid (CSF) circulation; Chiari Type I malformations, meningitis, basal arachnoiditis, cervical spondylosis, hydrocephalus, and posterior fossa arachnoid cysts.^[9] They are characterized by spinal cord enlargement and reversible parenchymal T1/T2 MR signal prolongation without cavitation.^[4] In general, they are asymptomatic or contribute to just mild myelopathy.

Syrinx formation attributed to ECF accumulation

Syrinxes occur due to disturbed ECF circulation/accumulation in the spinal cord attributed to scarring of the arachnoid and traction of the spinal cord (i.e., the dorsal cord is most susceptible because of its anatomy).^[1,3,5-8] These pathological changes disturb the absorption of ECF and contribute to the accumulation of ECF in the posterior gray matter, which eventually result in mature syrinx formation.

CONCLUSION

Chronological imaging from birth to 10 years of age showed progression of a “presyrinx” to a mature syrinx in a patient who as an infant presented with a lipomyelomeningocele and tethered cord.

Declaration of patient consent

The Institutional Review Board (IRB) permission obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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