Presyrinx state


Hydrocephalus is known to be associated with syringomyelia; however, the association between arrested hydrocephalus and syringomyelia has been reported only once before. This presyrinx state is a recently recognized condition that is known to resolve with proper treatment. This case report highlights a rare association between arrested hydrocephalus and the presyrinx state and outlines the implications for treatment and etiopathogenesis.


We agree with the authors that this is an extremely rare imaging finding in a child with presumed arrested hydrocephalus. However, we question whether the patient really had arrested hydrocephalus. The computed tomography scan of the brain revealed periventricular lucency, a finding that suggests that the hydrocephalic state had decompensated, and the presence of the presyrinx state indicates a dynamic condition caused by active but unstable pathological entities.1–3

We would like to share our experience with the presyrinx state occurring in another clinical situation. A 10-year-old otherwise healthy girl presented after being generally unwell for 14 months. Her symptoms consisted of occasional headaches and intermittent vomiting and diarrhea for 6 to 7 months; 6 weeks prior to admission she began experiencing progressive gait unsteadiness, deterioration in her writing, and intermittent urinary incontinence. On examination, she was alert and oriented but somewhat slow to react to commands. Her head was large, her pupils reacted briskly, and she exhibited lateral gaze nystagmus. Fundoscopy revealed papilledema. She had bilateral upper- and lower-limb ataxia, with gait and truncal ataxia. The remainder of her neurological examination was normal. An MR imaging study showed a midline cystic tumor located in the superior vermis causing compression of the pons and medulla as well as obstructive hydrocephalus and displacement of the tonsils into the foramen magnum (Fig. 1). The signal intensity was low on T1-weighted and high on T2-weighted MR images of the cervical spine, consistent with what is described as the presyrinx state (Fig. 2).1 A posterior fossa craniotomy and excision of the tumor were performed shortly after presentation. The tumor was found to be a pilocytic astrocytoma. The patient recovered from the surgery and did not suffer any new neurological deficits; her initial symptoms resolved after a few days. An MR imaging study was performed 1 month following tumor resection and revealed resolution of the presyrinx state in the cervical cord (Fig. 3) and no residual tumor.

The incidence of syringomyelia associated with tumor-induced tonsillar herniation has been reported to be as high as 21% in a study performed predominantly in adults.4 In the pediatric age group, however, the incidence is thought to be much lower. In our practice, in which approximately 20 children with posterior fossa tumor are examined annually, we have found that the incidence of tonsillar herniation is at least 60%; however, except for the patient presented in this letter we have not seen others with either a presyrinx state or syringomyelia in this setting.

The pathogenesis of both the presyrinx state and syringomyelia is unclear but may be due to altered cerebrospinal fluid (CSF) flow dynamics across the foramen magnum caused by impaction of the herniated cerebellar tonsils through the foramen magnum with obliteration of the subarachnoid space. The reason for the change in signal intensity of the cord, caudal to the subarachnoid obstruction is also not known, although it may reflect impaired transparenchymal fluid flow within the cord secondary to venous
obstruction at the level of the subarachnoid obstruction or differential venous pressures across the obstruction.

To our knowledge, the association of a presyrinx state with a posterior fossa tumor has not been reported. Other reported conditions found in association with the presyrinx state include Chiari malformation Type I,\textsuperscript{1} hydrocephalus following subarachnoid hemorrhage,\textsuperscript{1} cervical spondylosis,\textsuperscript{1} posterior fossa arachnoid cyst,\textsuperscript{2} and traumatic pseudo-meningocele formation.\textsuperscript{1} Relief of the subarachnoid obstruction, achieved in this patient by removal of the tumor, caused the imaging findings of a presyrinx state to reverse back to normal as reported by others.\textsuperscript{1,2}

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\textbf{References}

\textbf{Response}: I thank Drs. Gan and Cochrane for their interest in our work and their addition to the literature on the “presyrinx state” by adding one of their cases.

Drs. Gan and Cochrane believe that our patient did not have arrested hydrocephalus but rather that she had active hydrocephalus from the beginning. We disagree with this assessment. Our patient first presented to another institu-

\begin{figure}
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\includegraphics[width=\textwidth]{image1}
\caption{A T2-weighted MR image showing increased signal in the C2–7 region that is consistent with a presyrinx state.}
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\begin{figure}
\centering
\includegraphics[width=\textwidth]{image2}
\caption{Sagittal MR images revealing complete resection of the tumor, complete resolution of the tonsillar herniation, and the presyrinx state.}
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