A Twelve-Year-Old Girl with Chiari Malformation

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Presentation

- Initial symptoms were shortness of breath and cough that began the day of admission with associated headache and fatigue. She did not have fevers, chills, sore throat, chest pain, abdominal pain, nausea, vomiting, or diarrhea.

- The patient's medical history was unremarkable until 8 months prior when she began to experience changes in her voice and weight loss, dropping eight dress sizes. Upon further questioning, her family reported that she had a decreased appetite and would pick at her food. The patient then endorsed difficulty swallowing at times.

Physical Exam

Vitals: BP 131/82, HR 134, RR 36, O2 Sat 94% on 4L NC, Temp 98.2F

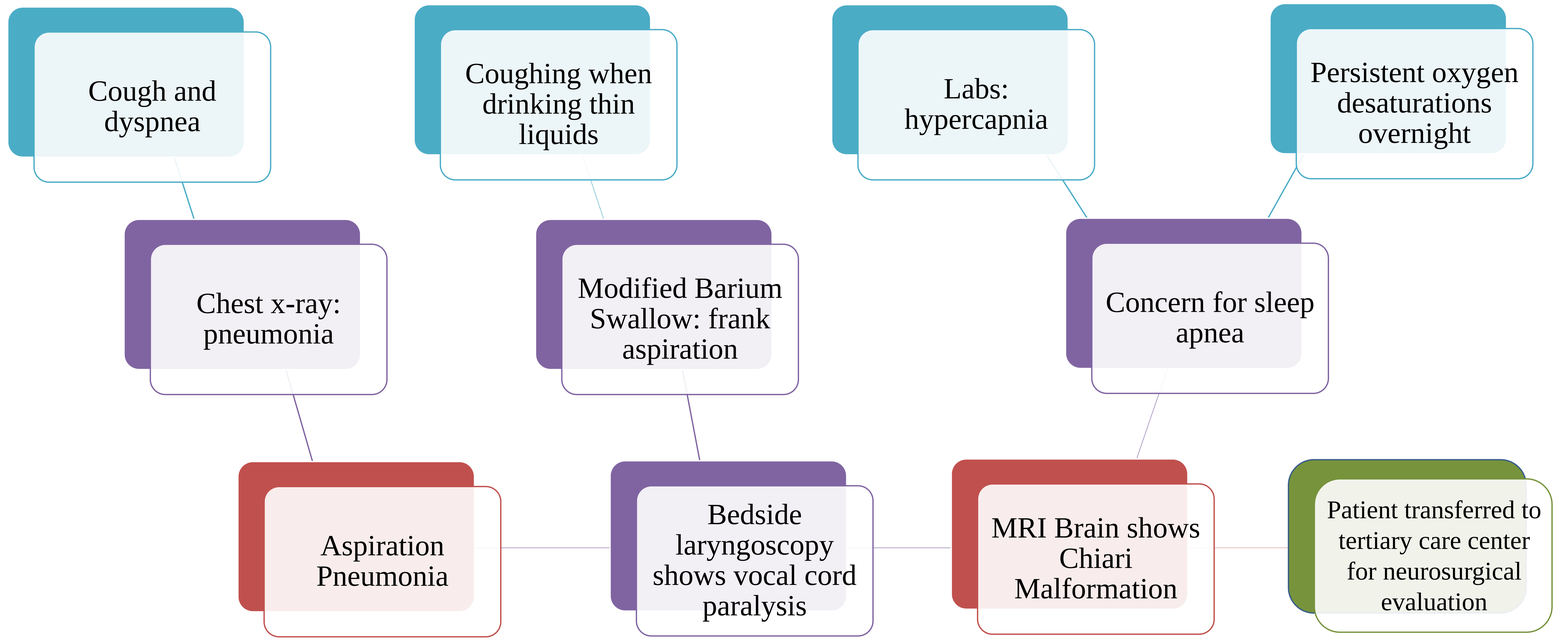
HEENT: Throat was clear without erythema, swelling, or exudates. Voice was breathy and she was unable to yell or speak loudly.

Respiratory: Chest wall was without retractions, but the patient was using her sternocleidomastoid muscles to breathe, especially while talking. Lungs were clear, but air movement was poor.

Cardiac: Tachycardic, regular rhythm.

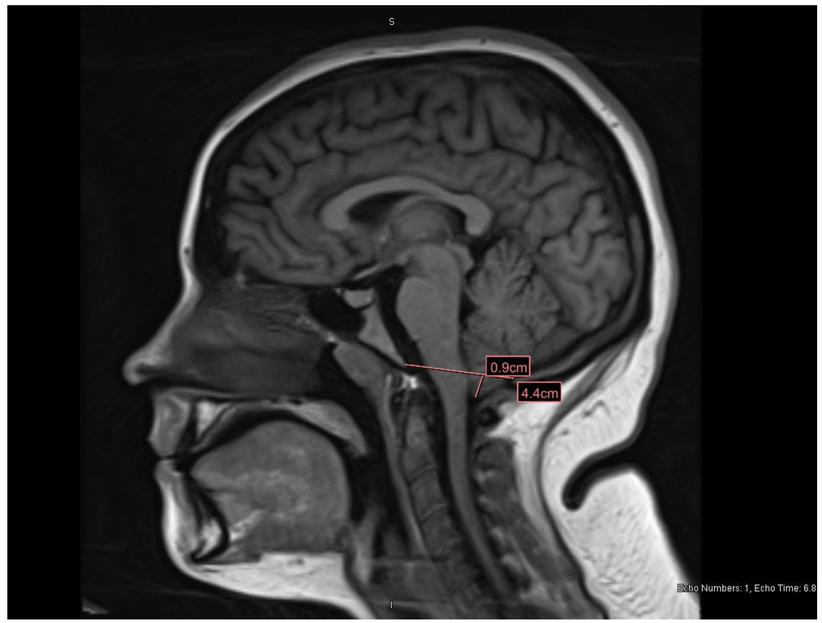
GI: Abdomen was obese, but nontender with positive bowel sounds.

Work Up



Discussion

- Chiari malformations, types 1-4, refer to a spectrum of congenital hindbrain abnormalities affecting the structural relationships between the cerebellum, brainstem, the upper cervical cord, and the bony cranial base.
- Classification is based on the morphology of the malformations. Chiari type I malformation is the most common and the least severe of the spectrum, often does not become symptomatic until adolescence or adulthood. It is characterized by abnormally shaped cerebellar tonsils that are displaced below the level of the foramen magnum.
- MRI is the most useful and most widely used imaging study for diagnosing Chiari malformation.
- Chiari I malformation is associated with the following symptoms: headache (most common), hoarseness, vocal cord paralysis, dysarthria, or sleep-related breathing disorders. Patients with Chiari I malformations who have minimal or equivocal symptoms without syringomyelia can be treated conservatively with clinical and MRI surveillance.
- Frankly symptomatic patients should be offered surgical treatment. The goals of surgical treatment are decompression of the cervicomedullary junction and restoration of normal CSF flow in the region of the foramen magnum.



References

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- Aspiration and Non-Aspiration Pneumonia in Hospitalized Children with Neurologic Impairment. Joanna Thomson, MD, MPH, et al. PEDIATRICS Volume 137, number 2, February 2016
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