

Chiari I Malformation

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ArnotHealth
Graduate Medical Education

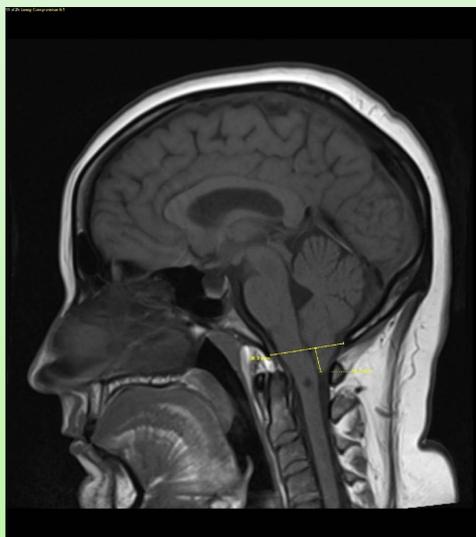
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INTRODUCTION

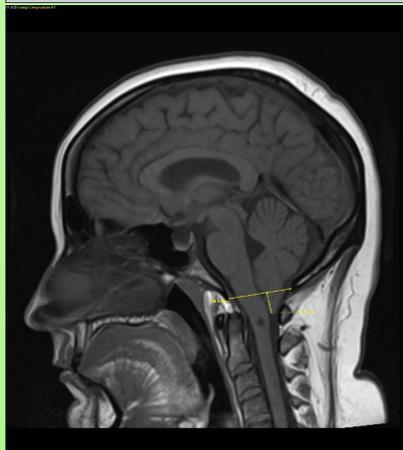
BACKGROUND: Chiari malformations are a spectrum of congenital anomalies of the cerebellum and posterior fossa primarily characterized by downward displacement of the cerebellar tonsils below the foramen magnum. Three types of Chiari malformations have been described with Chiari type I being the most common. Chiari type I is estimated to be present in 0.5-1% of the population and is asymptomatic in as many as 50% of individuals. Chiari I is characterized by downward displacement of the cerebellar tonsils at least 5 mm below the foramen magnum. CM 1 displays a strong association with hydrosyringomyelia, which is estimated to be present in 40-75% of cases. Symptoms include neck pain, occipital headache, weakness, spasticity and capelike upper extremity sensory loss. Pathogenesis is debated with multiple theories having been proposed. Research suggests it is not an inherited disorder. The etiology of syrinx formation is likely related to obstructed CSF flow at the foramen magnum. As a neuroanatomical disorder, imaging plays a crucial role in the diagnosis and treatment of CM 1, with MRI being preferred for its degree of anatomical detail. Tonsillar herniation due to elevated intracranial pressure should be ruled out before a diagnosis of chiari malformation is given. Surgical intervention is generally recommended in symptomatic patients with associated hydrosyringomyelia given its tendency to enlarge causing worsening symptoms and potentially permanent neurologic deficits. Improvement or stabilization of symptoms following surgery has been reported in approximately 94% of patients. This is a case of a symptomatic adult female, initially diagnosed with Chiari type I and syringomyelia who was imaged over the course of several years before and after surgical decompression.

Presentation

A 37 year old female with a history chiari I malformation diagnosed approximately 1 year earlier presented to her primary care physician with worsening headaches, tinnitus, visual changes, neck pain and upper back pain. An MRI was then obtained confirming low lying cerebellar tonsils approximately 1.1 cm below the foramen magnum as well as a 2mm syrinx. Medical management with Acetazolamide 250mg bid and NSAIDS were initially trialed. Follow up imaging demonstrated an enlarging syrinx measuring 5mm at 25 months and 9mm at 39 months following her initial presentation. Due to worsening symptoms and an enlarging syrinx, approximately 40 months following her initial presentation, the patient underwent posterior decompression surgery with C1 laminectomy and duraplasty at an outside facility. Approximately 9 months following surgery she reported occasional recurrent symptoms 1-2x per month consisting of intermittent tinnitus, vision changes, headache, upper extremity weakness as well as a general sponginess at her posterior head. Subsequent imaging showed that the syrinx was decreasing in size, measuring 3mm at 13 months and 2mm at 19 months following surgery.



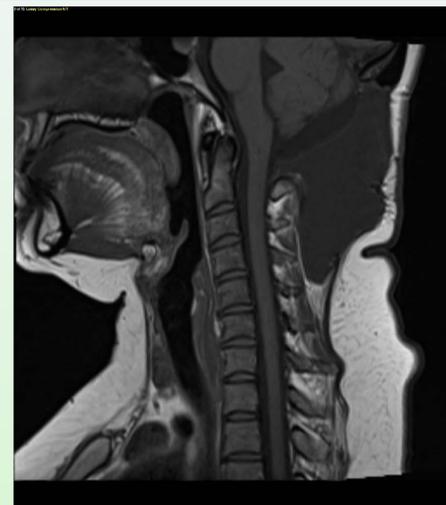
T1W sagittal MRI: Performed soon after initial presentation showing displacement of the cerebellar tonsils approximately 1.1 cm below the foreman magnum and a 2mm syrinx.



T1W sagittal MRI: Performed 9 months following initial presentation showing displacement of the cerebellar tonsils approximately 1.2 cm below the foreman magnum and an enlarging syrinx measuring 5mm.



T1W left and T2 right sagittal MRI images performed 39 months after initial presentation showing displacement of the cerebellar tonsils approximately 1.2 cm below the foreman magnum and a 8mm syrinx.



T1W sagittal MRI image Performed 13 months following surgical decompression showing a decrease in size of the syrinx which then measured 3mm.



T2W sagittal MRI image Performed 19 months following surgical decompression showing a decrease in size of the syrinx now measuring 2mm.

REFERENCES

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