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Case Report

Spontaneous Resolution of Aberrant Cerebellar Tonsil Movement in a Patient with Improving Chiari I Malformation☆

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ABSTRACT

Chiari malformation Type 1 (CMI) is traditionally characterized as a descent of the cerebellar tonsils more than 5mm below the foramen magnum. In some patients with CMI, there is aberrant pulsatile movement of the tonsils downward during cardiac systole which can affect cerebrospinal fluid (CSF) flow at the foramen magnum. Here, we present an 18-year-old female patient with CMI who presented with worsening symptoms of her CMI. Magnetic resonance imaging (MRI) at this time indicated an increase in cerebellar tonsil movement and decreased CSF flow at the foramen magnum. At her follow-up appointment, she had complete resolution of the aberrant motion of her tonsils and CSF flow returned to baseline without surgical intervention. Her symptoms also improved during this time, and she is now able to be followed by her primary care physician. The increased pulsatile movement of cerebellar tonsils in patients with CMI has been linked to diminished CSF flow at the foramen magnum and symptom severity. Spontaneous resolution of CMI is rare and has only ever been documented as ascension of the cerebellar tonsils. This case describes restoration of normal tonsil movement and baseline CSF flow corresponding with a resolution of symptoms where a complete resolution in tonsillar ectopia was not present.

Introduction

Chiari malformation Type 1 (CMI) is traditionally defined as a descent of the cerebellar tonsils more than 5mm below the foramen magnum [1]. Though not all patients with CMI experience symptoms, of those who do, patients can experience a vast array of symptoms including headache, neck ache, weakness, nystagmus, and apnea with headache being the most common (up to 50% of cases). Traditional management

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of symptomatic CMI is a posterior fossa decompression, however, there are a few cases reported in the literature of spontaneous resolution of tonsillar ectopia without surgical intervention [2,3].

Cerebellar tonsillar descent increases central nervous system (CNS) tissue motion and disrupts cerebrospinal fluid (CSF) flow dynamics at the cervicomedullary junction during the cardiac cycle [1,4–7]. In CMI, tonsillar motion is pulsatile because of pressure changes from the heart. This pulsatile motion results in disrupted CSF flow that many propose is associated with the severity of symptoms [1,7–9]. Here we present the case of a pediatric patient who had symptom relief following spontaneous resolution of aberrant CNS tissue motion and restoration of CSF flow.

Case presentation

A 9-year-old female with no previous medical history presented to the pediatric neurosurgery clinic for evaluation of a newly diagnosed CM1. She had a 3-month history of headaches located frontally and bilaterally with associated light and sound sensitivity and a 1-month history of bilateral eye twitching. Magnetic resonance imaging (MRI) showed cerebellar tonsils extending 5 mm inferior to the foremen magnum with no evidence of a syrinx. Due to the mild nature of her symptoms, she was monitored with follow up appointments and MRIs for worsening symptoms.

The patient's symptoms remained unremarkable until just over 6 years later when she returned to the neurosurgery clinic at age 15 with a 6-month history of worsening symptoms. She reported headaches located frontally and on top of her head with a pain rating of 8/10. The patient described episodes of "blacking out" after athletics in which her vision would go black, but she remained aware of what was going on around her. She also reported feeling weaker and slower in sports and intermittent instances where her feet and hands would go numb which did not coincide with the headaches. She also had her friends tell her that she snores. She was not taking any medications other than over the counter Ibuprofen which was ineffective in managing her headaches. Sagittal Fiesta Gated MRI sequence revealed cerebellar tonsils extending 8 mm inferior to the foremen magnum (Fig. 1). Sagittal Cine-Phase Contrast MRI revealed paucity of CSF flow dorsally and ventrally (Fig. 2), and abnormal, pulsatile neural movement at the level of the foremen magnum. At this time, she received a referral to ophthalmology to rule out papilledema and was asked to return to her primary care physician for treatment of her allergies before scheduling her for surgery. There was no evidence of papilledema so treatment for elevated intracranial pressure was not indicated. She was scheduled for follow up to discuss surgical options.

She returned for follow up approximately 1 year later at age 17. Her headaches were less often though still severe in nature, and she was still snoring at night; however, her other symptoms had resolved by this time. Due to relative decrease in symptom severity, it was decided to forgo surgery at this time. No imaging was obtained for this appointment. She returned 1 year later at age 18 for a follow-up MRI and

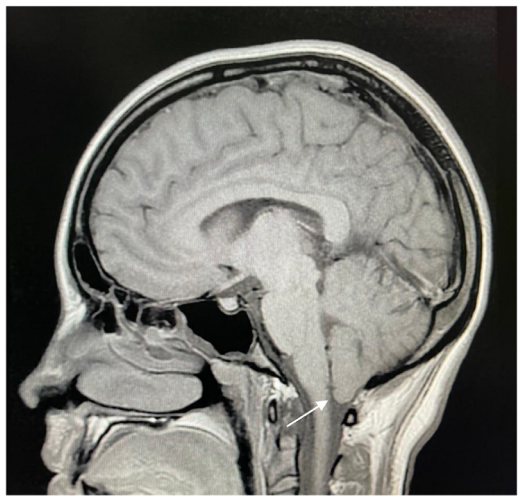

Fig. 1 – Sagittal T1 sequence MRI, demonstrating cerebellar tonsils extending below the level of the foramen magnum (white arrow).

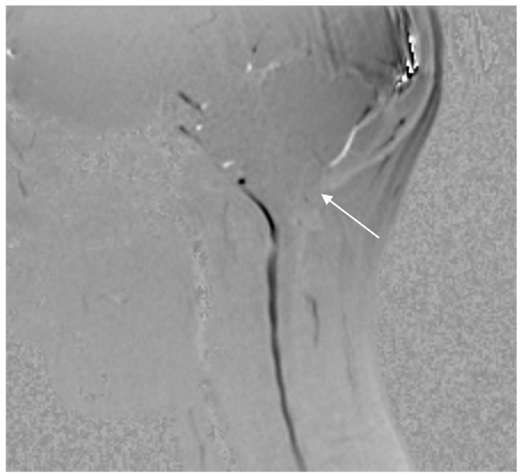

Fig. 2 – Sagittal cine-phase contrast MRI demonstrates decreased CSF flow dorsally at the level of the foramen magnum (white arrow).

appointment. MRI showed her cerebellar tonsils still extended 5 mm inferior to the foremen magnum (Fig. 3), however, the tonsillar shape was more rounded, rather than pinpoint as in the previous study. Chiari motion was less dynamic on the gated FIESTA MRI sequence. CSF flow at the foramen magnum had improved dorsally and ventrally (Fig. 4). She reported headaches were very rare with no other associated symptoms. She currently plays college sports without issues. It was decided that this patient was able to be followed as needed with her primary care physician.

Discussion

Spontaneous resolution of CMI is an uncommon phenomenon and very few cases have been reported in prior literature. Tra-

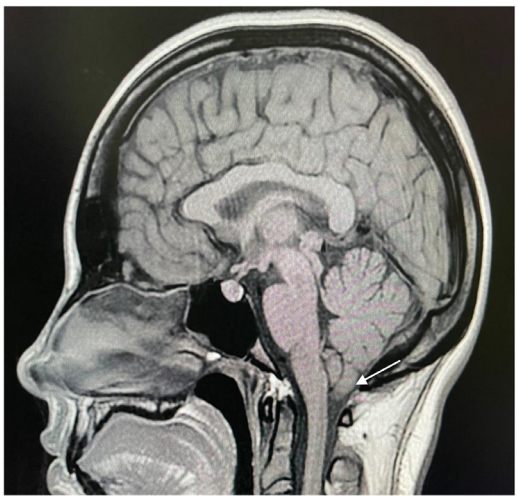

Fig. 3 – Sagittal T1 sequence MRI demonstrates improvement in the ectopia of the tonsils, in comparison to the previous study (white arrow).

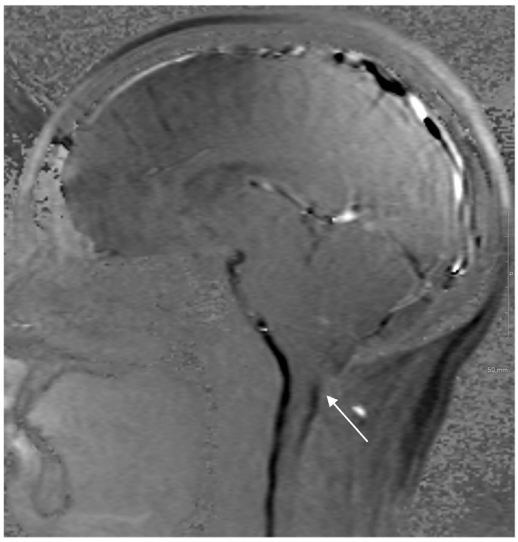

Fig. 4 – Sagittal cine-phase contrast MRI demonstrates improvement in CSF flow dorsally at the level of the foramen magnum (white arrow).

ditionally, spontaneous improvement of CMI has been seen in young children more often than adults as cerebellar tonsils normally ascend with age due to the increase in the posterior fossa volume. These cases resulted in resolution of tonsillar ectopia as posterior volume increased [3,10,11]. Only 5 instances of spontaneous resolution of CMI have been report in adult patients. In these patients there was significant hernia regression and symptom resolution [3]. Though spontaneous ascension of the cerebellar tonsils is more common in pediatric patients, symptom resolution is not always present. In a report by Waldau et al, they describe the case of a 3-year-old patient who had spontaneous resolution of a 13 mm CMI by age 7. Over the 4-year span, the patient had an 11% increase in posterior fossa expansion. This increase in growth of the posterior fossa was the expected rate of a child at this age and the volume of the posterior fossa was within normal size limits at both measurements [11]. In a similar case presented by Sun et al, a 7-year-old patient experienced complete resolution of his 13 mm CMI at the age of 13 after growing axially by 34 cm. Though there was resolution in the CMI, the patient still suffered from migraine-like headaches. The axial growth in this case is thought to correlate with continued growth of the cranial vault at the craniocervical junction thus pulling the cerebellum upwards. This mirrors previous reported cases in which cerebellar tonsillar descent resolved yet neurological symptoms persisted [10]. Unlike the patients mentioned above, our patient's tonsillar descent improved slightly, though she still met criteria for CMI. Most notably, there was complete resolution of the aberrant tonsil motion and CSF flow was restored when her symptoms improved. Furthermore, because she is in her late teens, cranial vault growth was not responsible for her improved tonsillar movement and CSF flow.

CNS tissue motion is generated during the cardiac cycle due to pressure changes resultant from pulsatile CSF flow [4,5,12]. The motion of the cerebellum, especially in the tonsils, can show an increase of up to 33% in patients in which CMI is present [12]. During cardiac systole, the cerebellar tonsils compensate for the increased pressure by further decent below the foramen magnum. The tonsils then return to their original position during diastole inhibiting the caudocranial flow of CSF at the level of the foramen magnum [13]. This can be visualized on cine-phase MRI as an increased flow velocity during diastole as well as decreased flow rate [4,9]. The altered CSF flow present in some patients with CMI is significant as previous cases have demonstrated a correlation between aberrant CSF flow and severity of symptoms. A study conducted by Hofkes et al detected abnormal CSF flow dynamics in symptomatic CMI patients compared to their non-symptomatic counterparts. This phenomenon is seen in our patient as her symptoms were at their worst when her tonsil movement and CSF flow was abnormal yet improved when the aberrant tonsil motion and CSF flow resolved.

Few prior studies report on the spontaneous improvement of cerebellar tonsil movement and resolution of CSF flow in the absence of complete regression of tonsil ectopia. As seen with this patient, her symptoms escalated when her tonsillar motion became more pronounced and CSF flow was diminished dorsally. Despite not receiving decompressive surgery, in her follow-up appointments, MRI showed no abnormal CNS tissue movement and return of normal CSF flow ventrally and dorsally at the level of the foramen magnum. Symptom improvement corresponded with resumption of normal CSF flow and resolution of marked tonsillar motion. This is a rare case of spontaneous improvement of aberrant tonsillar motion and CSF flow resulting in resolution of the patient's symptoms.

Patient consent statement

I certify that the patient presented here gave written consent for the publication of this case report and the images entailed within it.

REFERENCES

[1] Dawes BH, Lloyd RA, Rogers JM, Magnussen JS, Bilston LE, Stoodley MA. Cerebellar tissue strain in Chiari malformation with headache. World Neurosurg 2019;130:e74–81. doi:10.1016/j.wneu.2019.05.211.

[2] Leung V, Magnussen JS, Stoodley MA, Bilston LE. Cerebellar and hindbrain motion in Chiari malformation with and without syringomyelia. J Neurosurg Spine 2016;24(4):546–55. doi:10.3171/2015.8.Spine15325.

[3] Briganti F, Leone G, Briganti G, Orefice G, Caranci F, Maiuri F. Spontaneous resolution of Chiari type 1 malformation. Neuroradiol J 2013;26(3):304–9. doi:10.1177/197140091302600309.

[4] Pujol J, Roig C, Capdevila A , Pou A, Martí-Vilalta JL, Kulisevsky J, et al. Motion of the cerebellar tonsils in Chiari type I malformation studied by cine phase-contrast MRI. Neurology 1995;45(9):1746–53. doi:10.1212/wnl.45.9.1746.

[5] Pahlavian SH, Loth F, Luciano M, Oshinski J, Martin BA. Neural tissue motion impacts cerebrospinal fluid dynamics at the cervical medullary junction: a patient-specific moving-boundary computational model. Ann Biomed Eng 2015;43(12):2911–23. doi:10.1007/s10439-015-1355-y.

[6] Pinna G, Alessandrini F, Alfieri A, Rossi M, Bricolo A. Cerebrospinal fluid flow dynamics study in Chiari I malformation: implications for syrinx formation. Neurosurgical Focus 2000;8(3):1–8. doi:10.3171/foc.2000.8.3.3.

[7] Hofkes SK, Iskandar BJ, Turski PA, Gentry LR, McCue JB, Haughton VM. Differentiation between symptomatic Chiari I malformation and asymptomatic tonsilar ectopia by using cerebrospinal fluid flow imaging: initial estimate of imaging accuracy. Radiology 2007;245(2):532–40. doi:10.1148/radiol.2452061096.

[8] Lawrence BJ, Luciano M, Tew J, Ellenbogen R, Oshinski JN, Loth F, et al. Cardiac-related spinal cord tissue motion at the foramen magnum is increased in patients with type I Chiari malformation and decreases postdecompression surgery. World Neurosurg 2018;116:e298–307. doi:10.1016/j.wneu.2018.04.191.

[9] Sadique SI, Pandey P, Chaudhuri AK. Cerebrospinal fluid flowmetry in pediatric patients with chiari malformation I with surgical implications. World Neurosurg 2020;135:e83–6. doi:10.1016/j.wneu.2019.10.184.

[10] Sun PP, Harrop J, Sutton LN, Younkin D. Complete spontaneous resolution of childhood Chiari I malformation and associated syringomyelia. Pediatrics 2001;107(1):182–4. doi:10.1542/peds.107.1.182.

[11] Waldau B, Domeshek LF, Leigh FA , Lum KC, Fuchs HE, Marcus JR, et al. Spontaneous resolution of a 13-mm Chiari malformation Type I in relation to differential growth of the posterior fossa volume. J Neurosurg Pediatr 2009;3(2):110–14. doi:10.3171/2008.10.Peds08200.

[12] Cousins J, Haughton V. Motion of the cerebellar tonsils in the foramen magnum during the cardiac cycle. Am Jf Neuroradiol 2009;30(8):1587–8. doi:10.3174/ajnr.A1507.

[13] Menick BJ. Phase-contrast magnetic resonance imaging of cerebrospinal fluid flow in the evaluation of patients with Chiari I malformation. Neurosurg Focus 2001;11(1):1–4. doi:10.3171/foc.2001.11.1.6.