

# Case Lessons 22

## Wait and Watch strategy in incidental asymptomatic Chiari I with syringomyelia and literature review

Stela Dodaj, Mirel Grada, Vojsava Leka, Eugen Enesi, Mentor Petrela

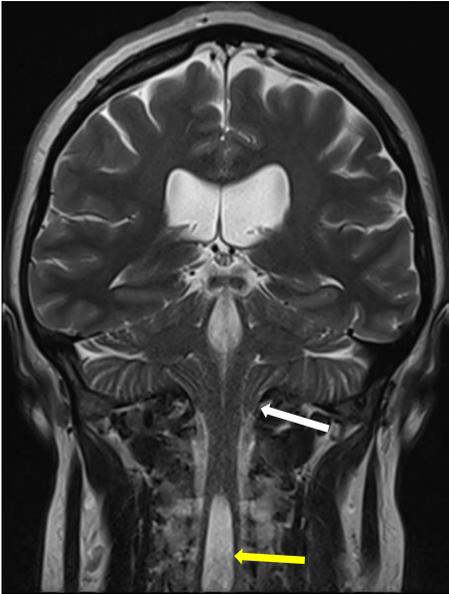
**Introduction:** Chiari malformation (CM), first described in 1891, is an anomaly of the posterior cranial fossa consisting of cerebellar tonsils descending through the foramen magnum. There are five types of CM from which CM type I (CMI) is the most common, and can be caused by a congenitally small size posterior fossa, or it can occur due to pressure changes such as high pressure above the cerebellum and low pressure below it. It is defined as herniation of cerebellar tonsils at least 5 mm below the foramen magnum. Whereas, types II-V, which escalate in severity of structures descending through foramen magnum and related symptoms, are congenital syndromes seen in children.<sup>1,2</sup>

Symptoms from CMI in adults usually appear before the fifth decade of life. The estimated proportion of adults with symptoms and radiological features of CMI is approximately 0.01-0.04%. The most common symptom is headache, however, there is a considerable number of patients without symptoms whose CMI is incidentally discovered in brain Magnetic Resonance Image (MRI) in co-existence with other pathologies.<sup>3</sup>

Decompression surgical treatment of posterior fossa aims to normalize CSF flow at the foramen magnum and reduce compression of the cervicomedullary junction. Asymptomatic CMI patients usually do not require surgery if there is no evidence of associated syrinx, while cases where syrinx is present are a matter of debate if surgery should be performed or not.<sup>4</sup>

**Case presentation:** We report a 34-years-old female who was referred for a resistant pituitary neuroendocrine tumor (PitNET) with active galactorrhea. During her radiological re-evaluation in brain MRI, besides the micro PitNET, CMI with syringomyelia from C2 to T5 was found as a co-occurrence. There were no symptoms related to these incidental findings. Her neurological examination was normal. Motor function and trunk sensibility were checked by two independent neurologists with the same results, nothing to be mentioned.

Both Somatosensory (SEPs) and Motor Evoked Potentials (MEPs) were performed and revealed no abnormality. Polysomnography was with normal result. Regarding the fact that there were no signs of compression at the cervicomedullary junction and this CMI patient had been asymptomatic, as well as motor and sensor tracts were unaffected by the associated syrinx, it was decided to wait and watch with annual follow-up evaluations.



**Fig. 1** MRI showing tonsillar herniation (white arrow) and syringomyelia (yellow arrow) on coronal T2-weighted sequence



**Fig. 2** MRI showing CMI (white arrow) and associated syringomyelia from C2 to T5 (yellow arrow) on sagittal T2-weighted sequence



**Fig. 3** Whole spine MRI showing syringomyelia from C2 to T5 (yellow arrow) on sagittal T2-weighted sequence

**Discussion:** The real incidence of clinically relevant CMI is considerably lower, despite the fact that the number of patients with the radiological diagnostic of CM has significantly grown since MRI became available. According to MRI studies, the prevalence has been shown to be between 0.56% and 0.77%, with a female to male ratio about 3:1.<sup>5</sup> Additionally, among adults, women are more likely to be diagnosed than men, and this contrast in sex is thought to be due to anatomical differences of anterior Cerebrospinal fluid (CSF) space (from the foramen magnum to the bottom of C2) between them, which is strongly associated with the symptoms of headache and neck pain. Thus, women, whose anterior CSF space is smaller, will show symptoms faster and more often than men.<sup>6-8</sup> In support of these findings, Garcia et al. also reported that head and neck pain increased with age in adult females CMI patients.<sup>7</sup>

Headache is the most frequent symptom of CMI. It is typically occipital or suboccipital and worsens during Valsava-like activities. Paresthesia of feet and/or hands is also seen if there is a compression of sensory tracts. Less frequently but significant are also sleep apnea, blurred vision, hearing problems and tinnitus, nystagmus, dizziness, and swallowing difficulties. These symptoms were absent in our patient. In some cases of CMI, patients develop syringomyelia. The incidence of syringomyelia in adult patients with CMI has been estimated at 69%.<sup>2,9</sup> A convincing theory on how syrinx was proposed

out by Oldfield et al. who suggested that fluid enters the spinal cord through the pulsatile movement of the cerebellar tonsils pushing on a restricting spinal subarachnoid space.<sup>5</sup>

In individuals without symptoms in whom CMI may have been discovered by chance on imaging performed for other reasons, surgery is not necessary if there is no syrinx. In case of evident symptoms, decompression surgery will significantly improve them, particularly the headache. An overall 83% improvement rate is reported in CMI decompression surgery.<sup>10</sup> It is still debatable whether an asymptomatic CMI patient with associated syrinx should undergo surgery or not.

However, according to nowadays guidelines,<sup>4</sup> it is suggested a close follow up in CMI patients with associated syrinx, and in case of syrinx expansion or symptoms appearing, surgery is the next step. The current treatment strategy is controversial because there are reported cases of spontaneous resolution of syrinx associated with CMI, when surgery is the generally accepted treatment modality.<sup>11</sup>

**Conclusion:** In case of incidental asymptomatic CMI with syringomyelia, it should be considered the possibility of spontaneous resolution over time. Attentive observation with periodic neurological and radiological evaluation should be done. Our case emphasizes the fact that wait-and-watch supports this approach.

#### References:

1. Friedlander RM. Congenital and Acquired Chiari Syndrome. *N Engl J Med*. 2024;390(23):2191-2198. doi:10.1056/NEJMra2308055
2. Arnautovic A, Splavski B, Boop FA, Arnautovic KI. Pediatric and adult Chiari malformation Type I surgical series 1965-2013: a review of demographics, operative treatment, and outcomes. *J Neurosurg Pediatr*. 2015;15(2):161-177. doi:10.3171/2014.10.PEDS14295
3. Heiss JD, Argersinger DP. Epidemiology of Chiari I Malformation. In: Tubbs RS, Turgut M, Oakes WJ, eds. *The Chiari Malformations*. Springer International Publishing; 2020:263-274. doi:10.1007/978-3-030-44862-2\_21
4. Pattisapu JV, Ackerman LL, Infinger LK, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Surgical Interventions. *Neurosurgery*. 2023;93(4):731-735. doi:10.1227/neu.0000000000002635
5. Holly LT, Batzdorf U. Chiari malformation and syringomyelia. *J Neurosurg Spine*. 2019;31(5):619-628. doi:10.3171/2019.7.SPINE181139
6. Fischbein R, Saling JR, Marty P, et al. Patient-reported Chiari malformation type I symptoms and diagnostic experiences: a report from the national Conquer Chiari Patient Registry database. *Neurol Sci*. 2015;36(9):1617-1624. doi:10.1007/s10072-015-2219-9
7. Allen PA, Loth F, Loth D, et al. Correlation of anterior CSF space in the cervical spine with Chicago Chiari Outcome Scale score in adult females. *Journal of Neurosurgery: Spine*. Published online December 1, 2024:1-10. doi:10.3171/2024.7.SPINE24370

8. Goel A, Jadhav D, Shah A, et al. Chiari 1 Formation Redefined—Clinical and Radiographic Observations in 388 Surgically Treated Patients. *World Neurosurgery*. 2020;141:e921-e934. doi:10.1016/j.wneu.2020.06.076
9. Ciaramitaro P, Massimi L, Bertuccio A, et al. Diagnosis and treatment of Chiari malformation and syringomyelia in adults: international consensus document. *Neurol Sci*. 2022;43(2):1327-1342. doi:10.1007/s10072-021-05347-3
10. Klekamp J. Surgical treatment of Chiari I malformation--analysis of intraoperative findings, complications, and outcome for 371 foramen magnum decompressions. *Neurosurgery*. 2012;71(2):365-380; discussion 380. doi:10.1227/NEU.0b013e31825c3426
11. Klekamp J, Iaconetta G, Samii M. Spontaneous resolution of Chiari I malformation and syringomyelia: case report and review of the literature. *Neurosurgery*. 2001;48(3):664-667. doi:10.1097/00006123-200103000-00044