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An Extension Form of Chiari-1 Malformation: A Case Report

Komang Yunita Wiryaning Putri[1,2*)] Novan Krisno Adji[1,3], Ulfa Elfiah[1,4] Muhammad Yuda Nugraha[1]

[1] Faculty of Medicine, University of Jember, Jember, Indonesia
[2]Department of Neurology, Soebandi Regional Hospital, Jember, Indonesia
[3] Department of Neurosurgery, Soebandi Regional Hospital, Jember, Indonesia
[4] Department of Plastic Surgery, Soebandi Regional Hospital, Jember, Indonesia

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*) *Corresponding author:*
E-mail: komangyunita.fk@unej.ac.id

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Abstract

The updated classification of Chiari malformation includes Chiari 1.5 malformation, which explicitly denotes the tonsillar herniation characteristic of Chiari 1 malformation, accompanied by an extended brainstem and a dilated fourth ventricle. We present a Chiari 1 malformation characterized by significant downward herniation of the tonsils and the development of syringomyelia, with no indication of elongation of the fourth ventricle. This example represents an extension of Chiari 1 malformation, which is currently not classified as Chiari 1.5 malformation. A 27-year-old female exhibited cephalic and cervical discomfort for two years. The initial MRI indicated a fall of the cerebellar tonsil beneath the foramen magnum. The midsagittal MRI assessed the degree of tonsillar herniation beneath the foramen magnum. Tonsillar herniation was observed at the C2 level; a spinal MRI revealed syringomyelia affecting the C2 to C3 levels. The patient received a suboccipital craniectomy and C1-C2 laminectomy. There was dural sac pulsation at the C2 level. The patient's cervical discomfort resolved entirely following the surgical procedure. This case represents an extension of Chiari 1 malformation, which is currently not classified as adenotes explicitly 1.5 malformation.

Keywords: Case report, Chiari malformation, Syringomyelia

Introduction

Chiari malformations (CM) are categorized into four kinds according to the extent of clinical and structural hindbrain involvement (Abd-El-Barr et al., 2014). Chiari I malformation is the inferior displacement of the cerebellar tonsils through the foramen magnum. The novel categorization of Chiari malformation is termed Chiari 1.5 malformation, which explicitly denotes the tonsillar herniation observed in Chiari 1 malformation, accompanied by an enlarged brainstem and fourth ventricle (Cools et al., 2023; Malik et al., 2015). The precise incidence of Chiari 1.5 malformation remains undetermined. Chiari 1.5 malformation is a rarer variant compared to Chiari 1 malformation. We present a Chiari 1 malformation characterized by significant downward herniation of the tonsils and the development of syringomyelia, with no indication of elongation of the fourth ventricle. This instance represents an expansion of Chiari 1 malformation, which has not yet qualified for the new classification of Chiari 1.5 malformation.

Case Presentation

A 27-year-old female exhibited cephalic and cervical discomfort for two years. She reported periodic cervical pain, primarily exacerbated by physical activity. She had no symptoms, including snoring, sleep apnea, weakness, or paresthesia, other than neck pain. The pain intensified with neck extension. She exhibited no prior symptoms or medical history of cerebral lesions, trauma, or any invasive procedures involving the brain or spine, including lumbar puncture.

The preliminary MRI indicated a fall of the cerebellar tonsil beneath the foramen magnum. The midsagittal MRI assessed the degree of tonsillar herniation beneath the foramen magnum. Tonsillar herniation extended to the C2 level; a spinal MRI revealed syringomyelia affecting the C2 to C3 levels. No sign of brainstem herniation, ventriculomegaly, or elongation of the fourth ventricle was observed (Figure 1).

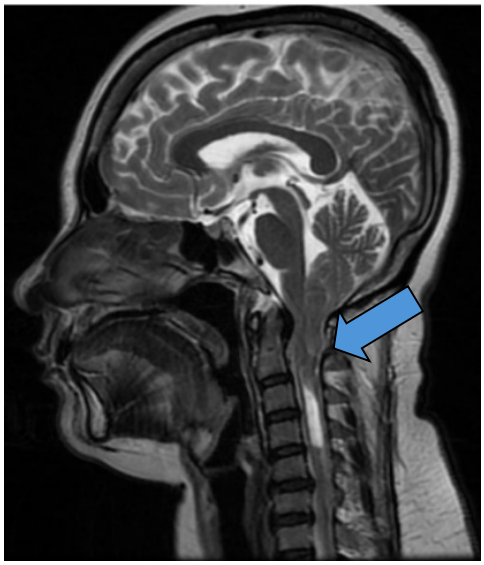

Figure 1. Midsagittal brain MR scans depicting the cervicomedullary junction and upper cervical spine, revealing cerebellar tonsillar herniation at the C2 spinal level (blue arrow) and syringomyelia.

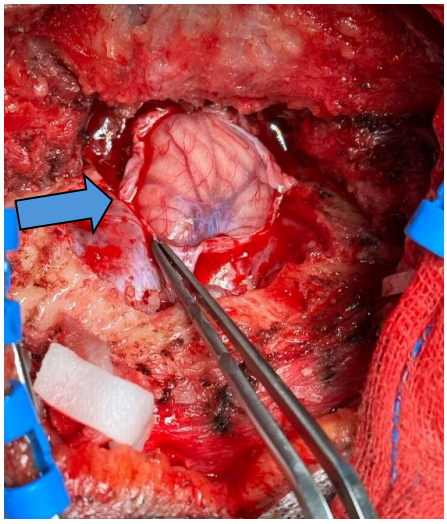

Figure 2. Intraoperative photograph of the cervicomedullary junction revealed cerebellar tonsils herniation (blue arrow) extending to the posterior arch of the atlas.

The patient received a suboccipital craniectomy and a laminectomy of the C1 and C2 vertebrae. The craniovertebral junction exhibited tightness, and dural sac pulsation was observable up to the C2 level (Supplementary video). The cerebellar tonsils exhibited symmetrical herniation (Figure 2). The tonsillar herniation extended below the foramen magnum to the C2 level. The dorsal aspect of the medulla oblongata appeared protruded, as if an external force had displaced it posteriorly. The obex and medulla were enlarged and relocated caudally to the lower segment of the C2 level. Operative findings validated the diagnosis of Chiari 1 malformation with herniation extending to C2.

Discussion

The spinal cord is the sole significant central nervous system structure that typically traverses the foramen magnum (Arnautovic et al., 2023). The descent of the cerebellar tonsils below the foramen magnum characterizes Chiari malformation (CM). The CM traditionally encompasses four classes based on anatomical entities. Type 1 refers to the inferior displacement of the cerebellar tonsils through the foramen magnum into the cervical canal. Type 2 or Arnold-Chiari malformation entails the herniation of the brainstem and inferior cerebellum into the cervical spinal canal. The fourth ventricle is positioned caudally and extends beneath the foramen magnum (Shoja et al., 2018). Type 3 is defined by the herniation of posterior fossa contents in an occipital or high cervical encephalocele (Ivashchuk et al., 2015). Type 4 refers to cerebellar hypoplasia or aplasia (Cools et al., 2023; Azahraa Haddad et al., 2018).

Recently, Chiari 0 and 1.5 classifications have been delineated (Tubbs et al. 2001 and 2004). The fundamental distinction between Chiari malformation type 1 and type 1.5 is the occurrence of caudal descent of the brainstem in the latter, alongside tonsillar ectopia. Distinguishing between the two entities is crucial for adequate care, since Chiari 1.5 patients are more prone to necessitate extensive and intricate

procedures in addition to decompression. The principal neuroimaging characteristic of Chiari 1.5 is the displacement of the obex and cerebellar tonsils beneath the foramen magnum (Moore et al., 2014).

The current instance had significant tonsillar herniation at the C2 level, accompanied by syringomyelia. She exhibited cephalic and cervical discomfort. Notably, no evidence indicated herniation of the posterior brainstem (obex), ventriculomegaly, or extension of the fourth ventricle. This case involved a Chiari 1 malformation characterized by significant tonsillar herniation, with no signs of brainstem or ventricular involvement (Yan et al., 2016).

The clinical symptoms and symptomatic results were comparable between the two groups regarding the extent of tonsillar herniation (Chiari 1 malformation vs. Chiari 1.5 malformation: 7 mm vs. 12.7 mm) and the incidence of syringomyelia (58% vs. 50%) (Tubbs et al., 2003). Neuroimaging can reveal characteristic radiological features in Chiari 1.5 malformation. Nonetheless, achieving an accurate preoperative diagnosis might be challenging, since posterior fossa congestion may obscure the identification of posterior fossa features (McVige et al, 2015). The differential diagnosis between Chiari 1 malformation and Chiari 1.5 malformation is crucial due to variations in surgical approach and treatment efficacy (Lei et al., 2018). Upon confirmation of Chiari 1.5 malformation in the patient, an aggressive surgical approach is warranted, entailing adequate foramen magnum decompression, which includes tonsillectomy and careful fenestration of intradural arachnoid adhesions and membranes, to resolve syringomyelia and avert the necessity for subsequent surgeries (Greenberg et al, 2015).

Conclusion

In this case, the advanced type of Chiari 1 malformation should be suspected of being Chiari 1.5 malformation. Chiari 1 malformation may extend to the C2 level; nonetheless, it does not meet the requirements for classification as Chiari 1.5 malformation.

Conflict of Interest

No potential competing interest was reported by the authors

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N/A

Author contribution

All authors will take public responsibility for the content of the manuscript submitted to the Journal of Agromedicine. KYWP had drafted a concept and prepared the manuscript, which was revised and received final approval for publication. NKA had a data analyst, an interpreter, and the person in charge of data collection in the field. UE had compiled the research design, interpreted the data, and provided final approval of the article. MYN had data collection and prepare the manuscript.

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