

## Case Report

# Chiari 1.5 malformation: a case report and review of literature

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## ABSTRACT

Chiari 1.5 malformation, a rare neurological disorder, involves cerebellar tonsil herniation and slight brainstem descent through the foramen magnum. A 25-year-old woman presented with recurrent headaches exacerbated by coughing, straining, and difficulty in maintaining head posture despite normal motor functions and reflexes. MRI revealed a Chiari 1.5 malformation with a 13.5 mm cerebellar tonsil herniation, minor brainstem descent, and C2-C3 vertebrae fusion. Suboccipital craniectomy with foramen magnum decompression significantly alleviated her symptoms. This case emphasizes the importance of recognizing Chiari 1.5 malformation as a cause of headaches in young adults and supports surgical intervention to prevent neurological decline. Although the aetiology is unknown, theories suggest a pressure differential between the cranial and spinal subarachnoid spaces during fetal development. Early diagnosis and posterior fossa decompression are crucial for symptom management and outcome improvement in CHIARI 1.5.

**Keywords:** Chiari 1.5 malformation, Headaches, Cerebellar tonsils herniation, Brainstem descent

## INTRODUCTION

Arnold-Chiari malformations (CM), is a group of structural abnormalities involving the cerebellum, brainstem, and craniovertebral junction. After Hans Chiari and Julius Arnold, it has evolved into a distinct classification of neurological disorders with significant clinical implications.<sup>1</sup>

### History and overview

Arnold-Chiari's malformation has a rich historical background. Hans Chiari first described the condition in 1891, categorizing it into three types based on autopsy findings. Julius Arnold, a German anatomist, added detailed descriptions of hindbrain herniation in 1894, particularly Chiari II malformations. The term 'Arnold-Chiari malformation' was later coined in 1907 by Schwalbe and Gredig, Arnold's students, to honour both contributors. Advances in imaging and neurosurgical techniques throughout the 20th century have refined our understanding of these malformations, leading to the

recognition of Chiari 1.5 and improvements in management strategies.<sup>2</sup>

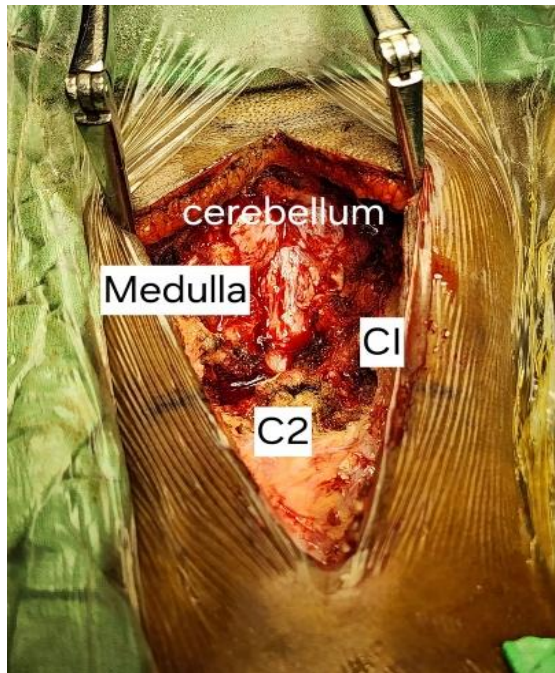
Chiari 1.5 malformation, an advanced form of Chiari I, includes brainstem descent and contributes to more severe clinical symptoms.<sup>3</sup> Despite the limited number of available case series, this malformation remains underexplored, warranting both case-specific insights and broader reviews of clinical and surgical outcomes.

## CASE REPORT

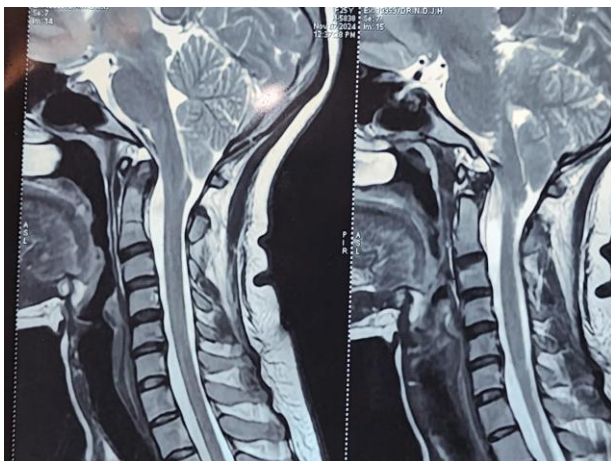
We encountered a 25-year-old female with recurrent occipital headaches for the past 2 years; however, last 2 months, she developed increased intensity headaches on stress, such as coughing, sneezing, and straining, and difficulty in holding her head straight. On general examination, the patient was found to be vitally stable.

Upon examination, the power of all limbs was 5/5. All deep tendon reflexes were normal. The cranial nerve examination results were normal, and no signs of

compressive myelopathy were observed. Gait was normal. Blood test results were normal.



**Figure 1: Intra op anatomy.**



**Figure 2: MRI brain and spine showing peg like herniation of cerebellar tonsil through foreman magnum and subtle flattening of the skull base.**

MRI brain and cervical spine was done and showed peg-like herniation of cerebellar tonsils through the foreman magnum by ~13.5 mm, and subtle caudal herniation of the brain stem through the foramen magnum. Crowding is seen in the Foreman Magnum. Obliteration of posterior CSF space in foreman magnum region. The fusion of C2 and C3 vertebral bodies/posterior elements is seen as a block vertebra. Subtle flattening of the skull base (measured ~145°) (platybasia). There was no evidence of syringohydromyelia or obvious dilatation of the ventricular system. These findings are suggestive of Chiari 1.5 malformation.

### **Management and outcome**

After anaesthesia evaluation, the patient underwent suboccipital craniectomy with foramen magnum decompression and C1 posterior arch excision under GA. The dura was not opened intraoperatively. Since we found the anatomy to be clear, she was electively intubated and weaned off the same evening. No further episodes of headache or neurological deficiency occurred during the postoperative period. Patient showed clinical improvement, discharged, and is currently on follow-up.

### **DISCUSSION**

The aetiology of Chiari 1.5 malformation is unknown, but one of the theories proposed for its pathogenesis involves the presence of a difference in pressure between the cranial and spinal subarachnoid spaces, which causes abnormal fetal brain development during pregnancy. A rare entity for headache which should not be missed when normal investigations do not reveal any definitive diagnosis of headache. A simple MRI of the brain and cervical spine is considered a non-invasive diagnostic modality employed to evaluate the brain, spinal cord, and CSF. MRI can be used to determine the extent of cerebellar herniation in the spinal canal. CM refers to a structural defect in the cerebellum, where a part of the brain tissue extends into the spinal canal.<sup>4</sup> It is classified into several types based on severity and anatomical abnormalities.

#### **CM type I**

*Description:* Cerebellar tonsils extend below the foramen magnum into the spinal canal.

*Symptoms:* Often asymptomatic but can include headaches, neck pain, balance issues, dizziness, muscle weakness, and problems with swallowing or breathing.

*Onset:* Usually diagnosed incidentally in adolescence or adulthood.

*Associated conditions:* Syringomyelia (fluid-filled cysts in the spinal cord) may develop.<sup>5</sup>

#### **CM type II**

*Description:* In addition to Arnold-Chiari malformation, both the cerebellar tonsils and brainstem extend into the spinal canal. The fourth ventricle may also herniate.

*Symptoms:* Symptoms appear during infancy and include breathing difficulties, swallowing problems, and arm weakness.

*Associated conditions:* Commonly associated with myelomeningocele (a form of spina bifida) and hydrocephalus (fluid build-up in the brain).<sup>6</sup>

### CM type III

*Description:* A severe and rare form in which a portion of the cerebellum and brainstem herniates into an encephalocele (sac-like protrusion) at the back of the skull.

*Symptoms:* Severe neurological deficits, including developmental delays and seizures.

*Prognosis:* Poor with significant complications.<sup>7</sup>

### CM type IV

*Description:* Involves underdevelopment (hypoplasia) or absence of parts of the cerebellum without significant herniation into the spinal canal.

*Symptoms:* Severe developmental and neurological problems, often fatal in infancy.<sup>8</sup>

### Other variants

*Acquired CM (Secondary Chiari):* Caused by conditions such as head trauma, infection, tumours, or excessive spinal fluid drainage.

*Complex CM:* Includes overlapping features or atypical presentations that do not fit neatly into the primary types.<sup>9</sup>

Essential difference between Chiari 1 and 1.5 is presence of caudal descent of the brain stem in the latter, along with tonsillar ectopia. These two clinical conditions share many similar features, and no signs/symptoms can differentiate between the two types of Chiari which are typically present in the second or third decade of life. However, young age at presentation and more severe symptoms, such as the bulbar sign, are more common in Chiari 1.5.<sup>10</sup>

### CONCLUSION

Chiari 1.5 malformation requires a high index of suspicion for diagnosis. Early surgical intervention is crucial for mitigating the progression of symptoms. This case adds to the growing body of evidence that supports posterior fossa decompression as a safe and effective treatment.

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*Ethical approval:* Not required

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