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Adult Onset Chronic Cephalous: A Rare Presentation of Arnold Chiari Malformation

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Abstract **Case Report**

The term Chiari malformation refers to a heterogeneous group of anatomical abnormalities at the craniovertebral junction. Chiari malformation type 1 (CM1) refers to the abnormal protrusion of cerebellar tonsils through the foramen magnum and is by far the commonest type. Its prevalence is estimated approximately 1%; it is more common in women and is associated with syringomyelia in 25-70% of cases. The prevalent pathophysiological theory proposes a morphological mismatch between a small posterior cranial fossa and a normally developed hindbrain that results in ectopia of the tonsils. In most people, CM1 is asymptomatic and diagnosed incidentally. In symptomatic cases, headache is the cardinal symptom. The typical headache is induced by Valsalva-like maneuvers. Many of the other symptoms are nonspecific, and in the absence of syringomyelia, the natural history is benign. The approach to patients with CM1 should be multidisciplinary. Magnetic resonance imaging, which shows cerebellar tonsillar decent 5 mm or more below the foramen magnum, is the gold standard investigative modality. Surgery is usually reserved for patients with disabling headaches or neurological deficits from the syrinx. Surgical decompression of the craniocervical junction is the most widely used procedure. Several surgical techniques have been proposed, but there is no consensus on the best treatment strategy, mainly due to lack of high-quality evidence. The management of the condition during pregnancy, restriction to lifestyle related to athletic activities, and the coexistence of hypermobility require special considerations.

Keywords: Cerebellar tonsils; Chiari malformation; Craniovertebral junction; MRI.

INTRODUCTION

Chiari malformation is a group of congenital deformities affecting the posterior fossa and hindbrain, characterized by the downward displacement of the cerebellar tonsils through the foramen magnum. The condition can present with a wide range of symptoms and signs, from mild to severe. This article discusses an adult presentation of type 1 Chiari malformation revealed through chronic cephalous, highlighting its clinical features, diagnosis, and management.

CLINICAL PRESENTATION

We report the case of a 24-year-old male, with the chief complaint of chronic headache located posteriorly aggravated while coughing or straining his neck and dizziness, with no history of fever, vomiting, fatigue, or photophobia.

Neural exam found no anomaly, and lab values were in the normal range.

A brain MRI was requested with the findings in favor of type 1 Chiari malformation.

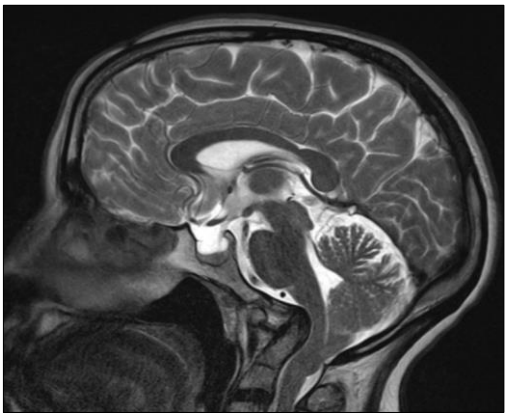

Fig. 1

DISCUSSION

The term Chiari malformation refers to a heterogeneous group of anatomical abnormalities at the craniovertebral junction. Chiari malformation type 1 (CM1) refers to the abnormal protrusion of cerebellar tonsils through the foramen magnum and is by far the commonest type. Its prevalence is estimated approximately 1%; it is more common in women and is associated with syringomyelia in 25-70% of cases. The prevalent pathophysiological theory proposes a morphological mismatch between a small posterior cranial fossa and a normally developed hindbrain that results in ectopia of the tonsils.In most people, CM1 is asymptomatic and diagnosed incidentally. In symptomatic cases, headache is the cardinal symptom. The exact pathophysiology of chronic cephalous in the context of Arnold Chiari malformation is not fully understood. However, it is believed that the downward displacement of the cerebellar tonsils through the foramen magnum can lead to compression of the brainstem and cerebellum, causing the symptoms observed in chronic cephalous.

The diagnosis of chronic cephalous is typically made through a combination of clinical evaluation, imaging studies, and laboratory tests. Imaging studies such as magnetic resonance imaging (MRI) are essential in confirming the presence of a Chiari malformation and assessing its severity. Laboratory tests may include cerebrospinal fluid (CSF) analysis to rule out other conditions that can cause similar symptoms.

The primary goal of treatment is to alleviate the symptoms and prevent further progression of the condition. This may involve surgical interventions such as decompression of the craniocervical junction to restore normal CSF flow and relieve pressure on the brainstem and cerebellum [1-3].

CONCLUSION

Chronic cephalous is a rare and debilitating presentation of Arnold Chiari malformation characterized by severe and persistent headaches. The condition is often associated with other symptoms such as dizziness, numbness, and difficulty with balance and coordination. Early diagnosis and management are crucial in preventing further progression and improving quality of life for patients with this condition.

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