Imaging of Chiari Type I Malformation and Syringohydromyelia

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KEYWORDS

• Chiari malformation • Syringohydromyelia • Syrinx • Headaches • Neuroimaging

KEY POINTS

- Chiari malformations are anatomic anomalies that comprise a broad spectrum of neurologic conditions.
- The most common malformation, a Chiari type I malformation (CMI), can present with a variety of signs and symptoms, most frequently an occipital Valsalva-induced headache.
- Cranial and spinal magnetic resonance (MR) imaging is used to identify the degree of tonsillar descent and document the presence of syringohydromyelia.
- Furthermore, this imaging serves as a baseline study that can be used for future comparison, particularly after a surgical procedure.
- The advent of cine-MR flow imaging (cine as in "cinema") has provided new insight as to the dynamic process involved in the evolution of this pathophysiology.
- It has also proved to be a useful tool to identify potential surgical candidates and evaluate postoperative progress.

Learning Objectives

1. Review the pathophysiology of Chiari malformations (CM) and syringohydromyelia (SH).
2. Recognize clinical signs and symptoms associated with CM and SH.
3. Radiographically define CM, SH, and associated pathologic condition.
4. Briefly discuss the medical and surgical treatments of CM and SH.
5. Understand the use of neuroimaging in preoperative and postoperative analysis of CM and SH.

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INTRODUCTION

The general term "Chiari malformation" (CM) refers to caudal displacement of the cerebellar tonsils through the foramen magnum; however, this is a heterogeneous disorder with varying degrees of pathology associated with a broad spectrum of signs and symptoms. The most common presentation is CMI; therefore, this is the primary focus of this article. The anatomy and pathophysiology of CMI and syringohydromyelia (SH), as well as the clinical signs and symptoms associated with these disorders, are presented. Radiological classification and diagnostic criteria of CMs and SH are reviewed. Finally, medical treatment and indications for surgery are also discussed.

BRIEF HISTORY

The diagnosis and treatment of CMI has been a topic of great debate in the literature since Austrian pathologist Hans Chiari (1851–1916) first discussed the disease at the end of the nineteenth century. Recent advances in neuroimaging and neurosurgery have provided new knowledge regarding the underlying pathophysiology, but the topic remains controversial because of the poor correlation between traditional neuroimaging modalities and clinical presentations.

The first case presented in Hans Chiari's initial work in 1891 described a 17-year-old girl who died of typhoid fever. She was found on autopsy to have "elongation of the [cerebellar] tonsils and medial divisions of the inferior lobules of the cerebellum into cone-shaped projections which accompany the medulla oblongata into the spinal canal." Chiari initially believed that herniation of the cerebellar tonsils was caused by chronic hydrocephalus.[1] This condition later became known as CMI.

Chiari subsequently described different but related conditions later termed Chiari types II (CMII) and III (CMIII). In further papers, he determined that the degree of hydrocephalus did not always correlate with the amount of cerebellar and spinal pathology. He hypothesized that poor growth of the bone in the posterior occipital region contributed to the likelihood of hindbrain herniation.[2] He cited both Cleland and Arnold in an 1896 publication discussing CMII.[2] Cleland[3] in 1883 had reported a case of an infant with hydrocephalus and spina bifida, and Arnold,[4] in 1894, a case of an infant with spina bifida and elongation of the cerebellar tonsils. Later, a series of cases of CMII malformations was reported by students of Arnold who referred to this as an "Arnold-Chiari malformation."[5] There was a great deal of confusion regarding the nomenclature, so, in 1971, Dreisen and Schmidt[6] proposed using only the term "Chiari malformation" to describe the type II presentation, which is currently the accepted terminology.

CLASSIFICATION

Diagnostic MR imaging criteria for CMI and CMI variants have been redefined by both age and associated pathology (**Box 1**, **Fig. 1**, **Table 1**). The presentation and severity of symptoms can change with age and time.[7] The measure of the cerebellar tonsils in normal individuals has been observed to decrease with age as measured on MR imaging by Mikulis and colleagues.[8] The measure can also increase with related intracranial pathologic changes such as hydrocephalus, mass lesion, and hypotension.

Imaging criteria for Chiari types II–IV are provided in **Box 1**, and representative images are shown in **Fig. 2**.