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SPECIAL ANNUAL ISSUE

Defining, diagnosing, clarifying, and classifying the Chiari I malformations

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Abstract

Purpose Chiari malformations (CM) have been traditionally classified into four categories: I, II, III, and IV. In light of more recent understandings, variations of the CM have required a modification of this classification.

Methods This article discusses the presentation, diagnostics, and treatment of the newer forms of hindbrain herniation associated with the CM type I.

Results The CM 1 is a spectrum that includes some patients who do not fall into the exact category of this entity.

Conclusions While CM have been categorically recognized as discrete and individual conditions, newer classifications such as CM 0 and CM 1.5 exhibit some degree of continuity with CM 1; however, they require distinct and separate classification as symptoms and treatments can vary among these clinical subtypes.

Keywords Chiari · Chiari 0 · Chiari 1.5 · Neurosurgery

Abbreviations
CM Chiari malformation
CSF Cerebrospinal fluid

Introduction

Chiari malformations are structural variations of the cerebellum and skull base and were first well described by Hans Chiari. These malformations have been classified as Chiari I, II, III, and IV malformations. However, some malformations do not fit neatly into any of these four categories as many of these differ anatomically or symptomatically. Chiari's original system recognized these malformations as specific, individual conditions. His nomenclature does not refer to tonsillar variations on a continuous spectrum [1]. As a result, additional subclassification is in order [2, 3].

According to Chiari's classification, malformations are grouped as discrete categories. Type I (Fig. 1) consists of a herniation of the cerebellar tonsil that extends inferiorly to the foramen magnum. However, in light of more recent advancements in clinical medicine, Chiari 0 (Figs. 2 and 3) and 1.5 malformations (Fig. 4) have been recognized as variations of the Chiari I malformation. Chiari 0 malformations are considered borderline defects in which a syringohydromyelia responds to decompression of the posterior cranial fossa although there is little to no cerebellar tonsillar herniation (< 3 mm). Chiari 1.5 malformations are more severe forms of Chiari I malformation in that more brain tissue crowds the foramen magnum. While symptoms and anatomical considerations are quite similar, treatment outcomes differ between Chiari I and 1.5 malformations. As a result, a thorough understanding of a newer classification system is crucial for the proper recognition and treatment of individuals with Chiari malformations that do not fit into traditional subtypes.

Chiari malformations

Chiari's description of the Chiari malformation type I (CM I) herniation was of cerebellar tonsils passing below the foramen magnum [3]. This was often associated with other anatomical variations of the skull and central nervous system [3, 4]. Though the exact underlying mechanism for hindbrain herniations is unknown, various theories have been proposed [5]. The initial description was made at autopsy and stated by

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Chiari as "elongation of the tonsils and medial divisions of the inferior lobules of the cerebellum into cone shaped projections

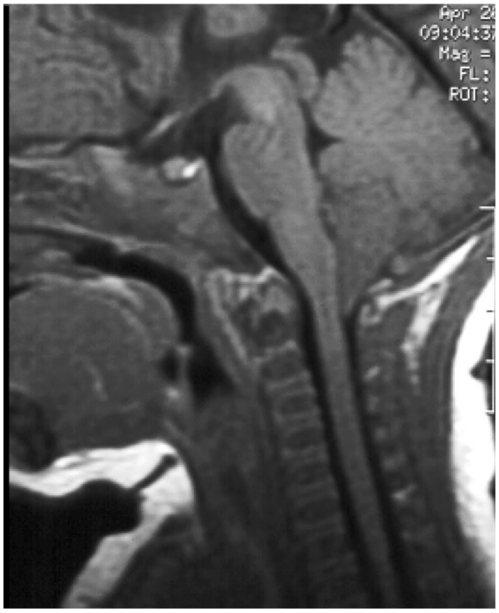

Fig. 1 Midsagittal, T1-weighted MRI of a patient with CM I

which accompany the medulla oblongata into the spinal canal" [6–8]. Chiari's nomenclature was limited in its description as it lacked a quantifiable amount of tissue herniation, though it did describe an asymmetric tonsillar ectopia [6]. Aboulezz et al. (1985) posited that within the normal population, the cerebellar tonsils may herniate up to 3 mm through the foramen magnum [9]. Barkovich et al. concluded that a minimal cutoff for the CM I would be 3 mm of tonsillar herniation [10]. It has been widely accepted that the inferior

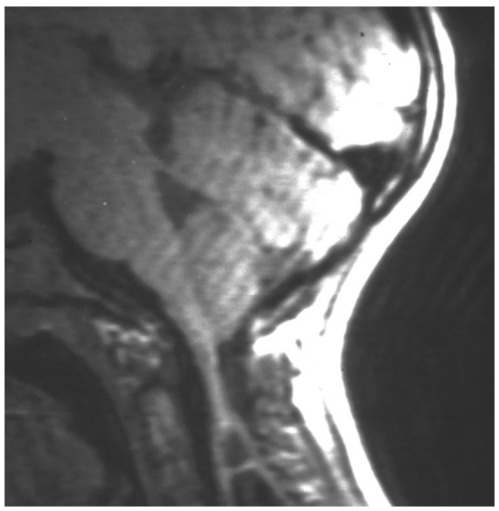

Fig. 2 Preoperative image of a patient with a large cervicothoracic syringomyelia and no hindbrain herniation. This cannot be classified as CM 0 until it is proven that the syrinx improves following posterior fossa decompression

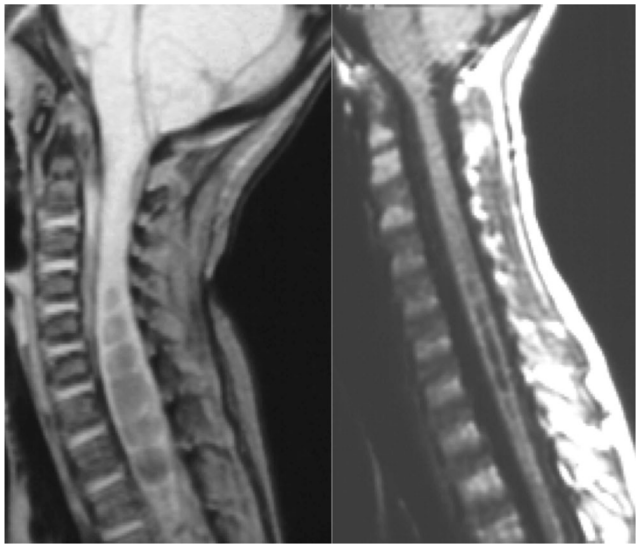

Fig. 3 Postoperative decompression of the craniocervical junction reveals a large preoperative syrinx (left) and no hindbrain herniation and a diminished postoperative syrinx (right) thus confirming the diagnosis of CM 0

cutoff for CM I herniations is 5 mm of tonsillar ectopia although interobserver differences in measurements of tonsillar herniation is well known [11]. CM I also has a variable presentation and findings requiring further classification. For example, in a study by Bindal et al., CM I was further subclassified into patients who were asymptomatic, experienced brain stem compression, and had syringomyelia. It was stated that this newer classification made descriptions

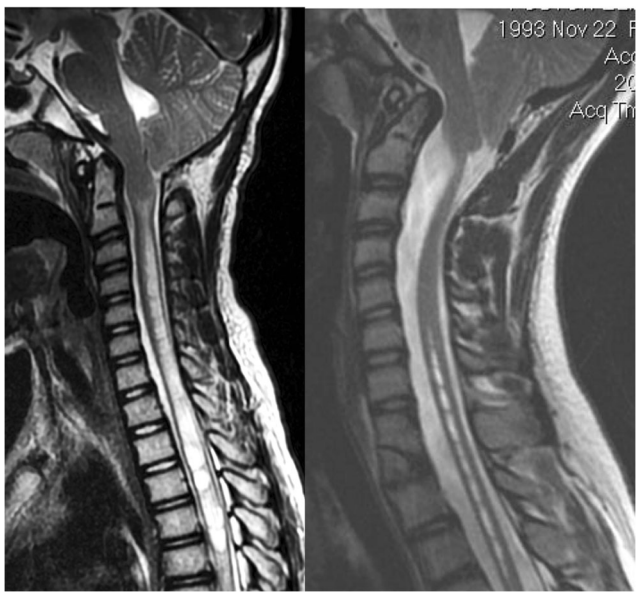

Fig. 4 Midsagittal, T1-weighted MRI of two patients with tonsillar ectopia consistent with CM I but with the addition of an elongated and caudally displaced brain stem thereby forming the so-called Chiari 1.5 malformation. Note the posteriorly displaced odontoid process in the patient on the right

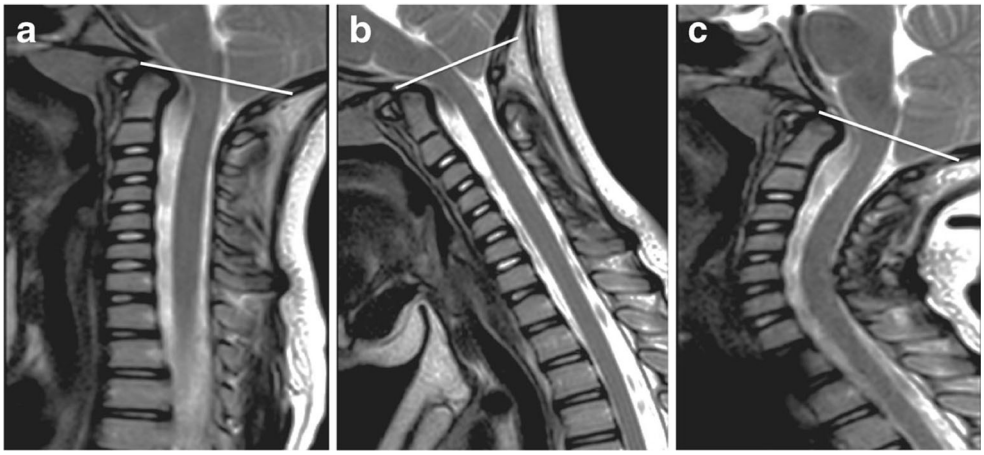

Fig. 5 Sequential images of a patient with CM I noting the position of the cerebellar tonsils in neutral (**a**), flexion (**b**), and extension (**c**). In this case, flexion results in the tonsils being located at a more inferior angle

more accurate, narrowed surgical options, and conveyed outcomes and long-term prognosis [12].

Diagnostics

Currently, though multiple planes are used to diagnose CM I, the gold standard is mid-sagittal MR imaging. This technique does have its limitations since the cerebellar tonsils are bilateral structures, the basion and opisthion are challenging to identify (especially in a younger population), evaluation of the degree of caudal descent is challenging postoperatively following removal of the opisthion, and flexion/extension of the head may change the location of the tonsils (Figs. 5 and 6) [13, 14]. Because the basion and opisthion on sagittal MRI are more challenging to locate in younger children due to underdeveloped intraosseous medullary spaces, there is an increased risk for overestimation of the amount of cerebellar tonsillar displacement.

Using coronal MR imaging, Tubbs et al. found asymmetry between the right and left cerebellar tonsils in 96% of patients, positing that CM I usually involves asymmetric tonsillar ectopia (Fig. 7) [15]. When studying a pediatric population showing symptoms related to CM I, 18% of the patients had clinical symptoms due to the unequal herniation. In addition, for those patients who also had a coexisting syringomyelia, 95% showed a greater amount of herniation of the right tonsil (Fig. 7). Furthermore, asymmetric tonsillar ectopia can lead to a misdiagnosis in the event midsagittal MR imaging captures the less descended tonsil while missing the contralateral tonsil.

In another study conducted by Tubbs et al., there was discrepancy between coronal and midsagittal planes when diagnosing CM I (Fig. 8) [15]. In most of the cases, coronal MRI did not meet criteria for CM I whereas midsagittal MRI qualified the patient for such a diagnosis. In addition, these studies found that one tonsil was often more caudally descended in coronal sections [15]. It was concluded that midsagittal MR imaging may overestimate CM I [15].

Treatment

According to Bindal, decompression surgery should be performed for all symptomatic CM I patients to alleviate symptoms and arrest progressive deterioration [12]. This surgical technique includes resection of the posterior foramen magnum

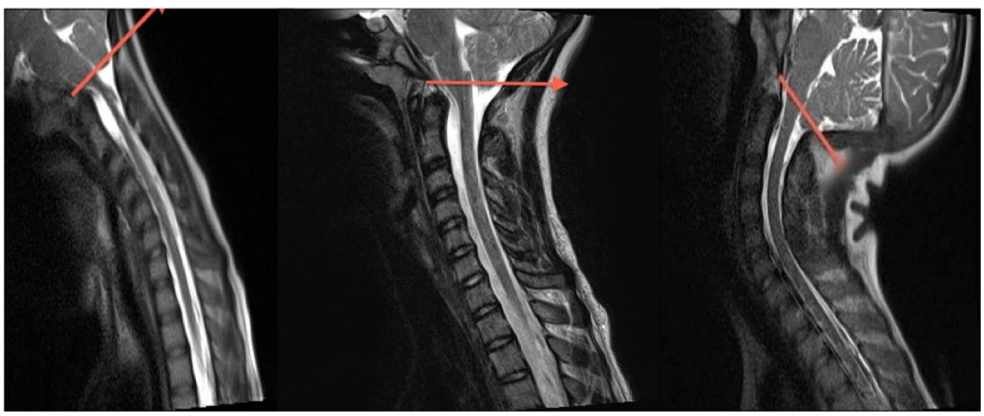

Fig. 6 Similar images as seen in Fig. 5 but in this patient with CM I, extension results in the tonsils being located at a more inferior level

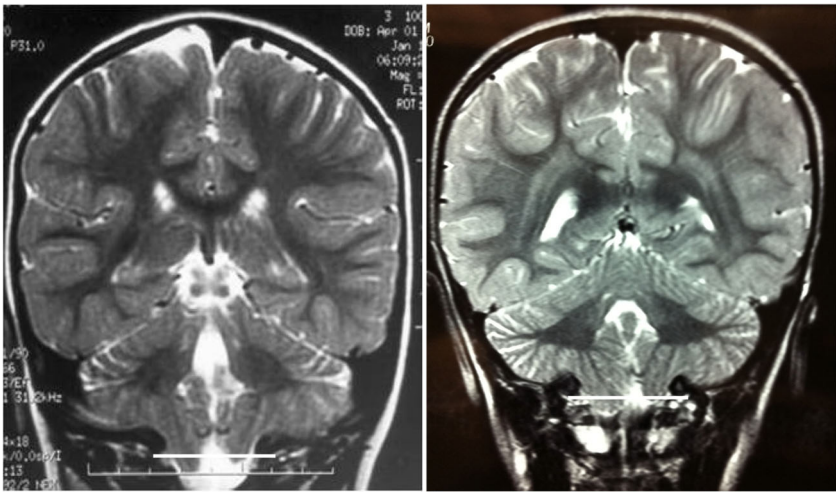

Fig. 7 Coronal images from two patients with CM I. Note on coronal imaging, the asymmetry between the left and right tonsils is seen and that in these two patients, the left tonsils alone would not qualify as CM I

and the posterior arch of C1, and ±duraplasty. There may also be indications for possible anterior decompression, especially with a retroverted odontoid process.

Chiari 0 malformation

Type 0 Chiari malformation (CM 0) is a subset of Chiari malformations in which patients have a syringomyelia with little to no associated hindbrain herniation (< 3 mm) that responds to craniocervical decompression (Figs. 2 and 3) [2, 4]. Similar to CM I, symptoms are thought to be precipitated by a craniocervical junction disruption to the flow of cerebrospinal fluid (CSF). This similarity has given CM 0 synonyms such as "borderline Chiari" or "Chiari-like pathophysiology" with a "tight cisterna magna" [16].

Originally termed syringohydromyelia without hindbrain herniation in 1969 by Newton, Iskandar et al. termed this condition CM 0 after analyzing cases in which the symptoms and syrinx size improved after posterior cranial fossa decompression [2, 17]. Although there have been a number of reported CM 0 cases since then, the overall number of case presentations has been few. CM 0 might also represent intermittent hindbrain herniation not seen on static imaging.

Features

Tubbs et al. completed a study of the posterior cranial fossa in patients with CM 0 and found deviations from controls [18]. Radiological imaging showed that the tips of the obices could be found in excess of two standard deviations beneath the mean value [18]. The spinomedullary junction midsagittal anteroposterior distance was increased at the level of the foramen magnum [18]. The angle between the base of the fourth ventricle and the clivus was elevated; however, the clival angles and prepontine spaces were found to be within normal limits [18]. Similar findings were also reported in other studies [19, 20].

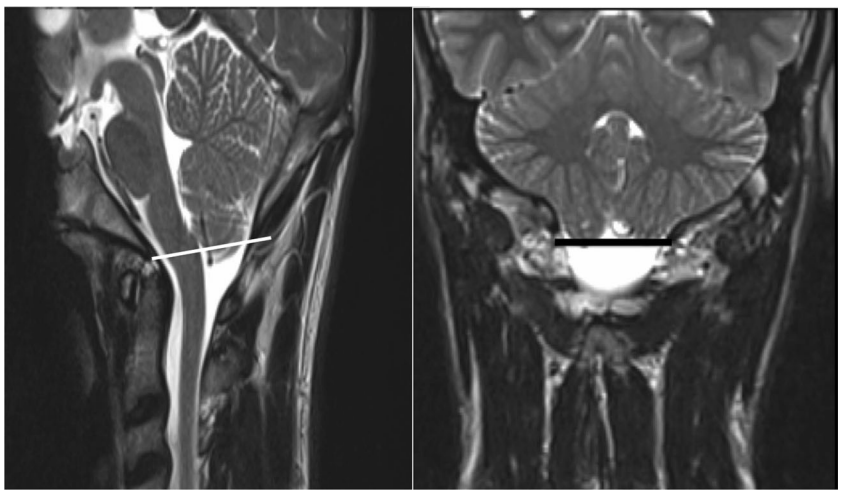

Fig. 8 A patient diagnosed with a minimal (3–4 mm) CM I via sagittal MR imaging (left) but with analysis of coronal MRI (right) no cerebellar tonsillar herniation is evident

In this population, Sekula et al. found that the angle of the tentorium cerebelli was greater, whereas the clivus, basisphenoid, and basioccipital were significantly shorter when compared with controls [21]. In addition, it was found that certain cases of syringomyelia lacking hindbrain herniation had impacted cerebellar tonsils in the plane of the foramen magnum with an open arachnoid space anterior to the brain stem [16].

Pathogenesis

In cases of a near virtual absence of cerebellar herniation as in CM 0, syrinx formation is most likely due to obstructed CSF circulation. This obstructed flow could be attributed to the veils or arachnoid adhesions in the fourth ventricle foramina that are often found during surgery. This usually involves the foramen of Magendie. CSF flow could also be obstructed as a result of a distorted posterior fossa [2, 16, 18].

Since there is more evidence of pathophysiological similarity between CM 0 and CM I, familial clustering suggests genetic as well as multifactorial factors, such as epigenetic and environmental causes, for the differentiation of the two [22]. Therefore, CM 0 and CM I may fall on a continuum rather than be distinct abnormalities.

Clinical presentation

CM 0 and CM I have similar reported presentations such as limb weakness, scoliosis, and paresthesias [23]. In addition, posterior Chiari-like headaches of short duration are typically present and can be induced by Valsalva maneuver [23]. These headaches are suggested to be symptoms of intermittently herniating tonsils, not seen on MR imaging [19].

Diagnosis and treatment

Though many techniques have been proposed to treat Chiari malformations, restoring normal CSF flow is required and performed via a posterior cranial fossa decompression [23, 24]. Badie et al. (1995) found that occipital craniectomy, with or without duraplasty, leads to cessation of symptoms postoperatively due to decreased craniocervical pressure dissociation [25]. This operation consists of removing a portion of the skull at the posterior aspect of the foramen magnum, removal of the posterior arch of the atlas, and ±incision of the dura, lysing the arachnoid adhesions/veils to re-establish normal CSF flow through the foramen of Magendie [2, 16]. This can be done with or without duraplasty.

Preoperatively, the diagnosis of CM 0 is one of exclusion and is only confirmed postoperatively with the improvement of clinical symptoms [23]. Finally, though this malformation has been used to describe tonsillar herniation of less than 3 mm in the absence of syringomyelia, this is not a proper use of the term [23].

Chiari malformation 1.5

Chiari malformation type 1.5 (CM 1.5) (Fig. 4) is a more complicated form of CM I with the tonsillar herniation seen in CM I but with the addition of lengthening and inferior displacement of the brain stem, and obex being located caudal to the basion-opisthion line [3, 26, 27]. Though the exact incidence of CM 1.5 is not known, it is proposed to be less common than CM I [28]. Since CM I and CM 1.5 share morphological and anatomical similarities, diagnostic distinction needs to be made because the operative outcomes can differ between them [26]. According to Tubbs et al., patients with CM 1.5 were more likely to fail the initial posterior fossa decompression compared with patients with CM I and usually manifested as continued presence of a syringomyelia [3, 29, 30].

Clinical presentation

The presentation of CM 1.5 is similar to CM I, yet no signs or symptoms differentiate the two. However, patients have reported headaches in the presence and absence of the Valsalva maneuver, shortness of breath, jaw pain, difficulty speaking, opisthotonos, absent gag reflex, lethargy, and drop attacks [3, 4, 23, 27].

Treatment

Treating CM 1.5 first depends on determination of the root cause of the pathology, including hydrocephalus,

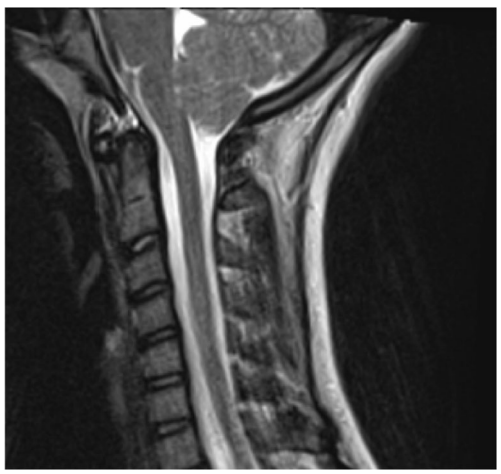

Fig. 9 Minimal descent of the cerebellar tonsils but in this patient, there are no signs of compression and these structures have a normal rounded appearance

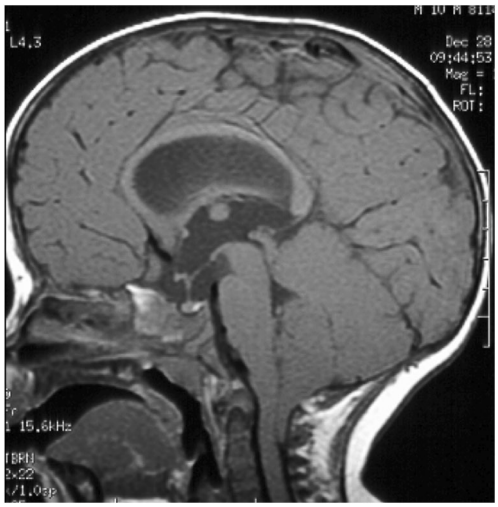

Fig. 10 Ventriculomegaly is an asymptomatic patient. Note the caudal displacement of the cerebellar tonsil that to some qualifies as a CM I but to others is simply a result of the enlarged ventricles

craniosynostosis, and/or a brain tumor. For patients with craniosynostosis, posterior occipital decompression is recommended [3, 31–33]. In addition, determination as to whether a syrinx is present should be made. If present, decompressive surgery is recommended to decrease the compression of the spinomedullary junction.

As with other Chiari malformations, posterior decompressive surgery is the only effective treatment; however, syringosubarachnoid shunting has been reported in treatment of CM 1.5 [3, 34]. In many cases of symptomatic CM 1.5 with inferior displacement of the brain stem, patients have a poorer response to posterior cranial fossa decompression, more so if they have concomitant syringomyelia [3, 29, 30].

Other

Some hindbrain herniations are not clear. Most often, a compressed and tapered cerebellar tonsil greater than 3 mm inferior to the foramen magnum is considered a CM I. However, is a rounded, normal appearing cerebellar tonsil with minimal caudal displacement (Fig. 9) in an asymptomatic patient also a CM I? Such patients with a rounded tonsil at or below the foramen magnum might be misinterpreted as a CM 0, which as previously stated must have syringomyelia that responds to posterior cranial fossa decompression. Additionally, are cerebellar tonsils that are found below the foramen magnum in the presence of ventriculomegaly or hydrocephalus to be considered CM I (Fig. 10)? Some have argued that the hydrocephalus preceeds the hindbrain herniation thus the hindbrain hernia is secondary and should not be considered in the spectrum of CM I. Lastly, some forme fruste of CM I might not be adequately classified or diagnosed as they fall outside of the "normal" presentations. For example, a stenotic foramen magnum might result in cerebellar tonsils that are herniated inferior to the foramen magnum but not posterior to the brain stem/ spinal cord as is usually seen. In these cases, the typical midsagittal imaging (see above) might miss this anatomical variant (Fig. 11), which should be considered in the spectrum of CM I.

Conclusion

Knowledge of the newer Chiari malformation classifications is required to improve patient treatment and outcomes. An improved understanding of their embryology and clinical manifestations will help in better categorizing these entities [35–37]. Although midsagittal MR

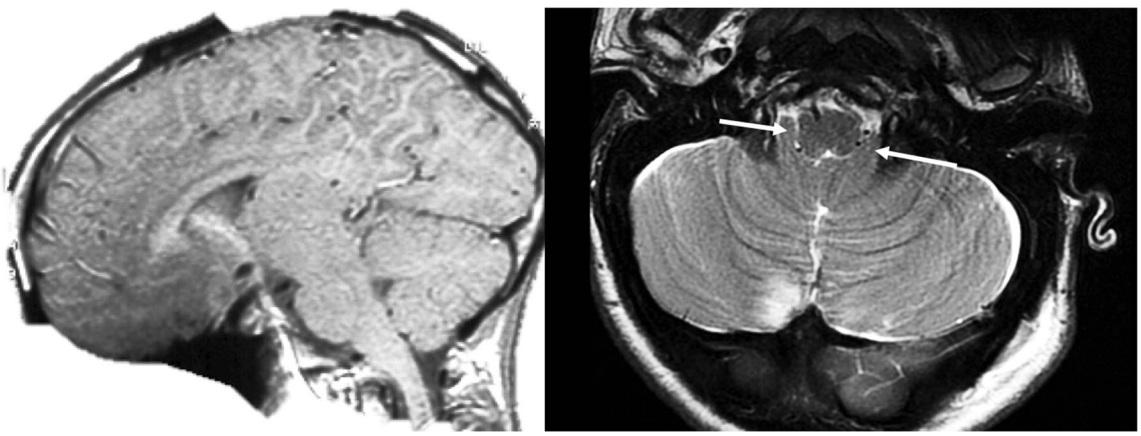

Fig. 11 A patient where no tonsillar ectopia is seen on midsagittal imaging (left) but with axial imaging (right) the cerebellar tonsils are seen lateral to the spinomedullary junction

imaging is widely accepted as a diagnostic standard, we highlight that such imaging may overestimate the degree of tonsillar ectopia and concomitant use of coronal MR imaging with measurements of both tonsils allow for a more accurate diagnosis of Chiari malformations.

Compliance with ethical standards

Conflict of interest None.

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