Clinical Manifestations of Chiari I Malformation

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KEYWORDS

• Chiari 1 malformation • Syringomyelia • Syrinx • Scoliosis • Headache • Cervicomedullary junction • Craniocervical junction • Oropharyngeal dysfunction

KEY POINTS

- Asymptomatic Chiari 1 malformation (CM1) patients often remain so over time. Those with mild symptoms are likely to remain stable and may improve.
- The most common clinical presentation of CM1 in children involves headaches, syringomyelia, and/or scoliosis.
- The most common headache associated with CM1 are occipital/suboccipital headaches, worsened by Valsalva-like maneuvers, and lasting less than 5 minutes.
- Oropharyngeal dysfunction is one of the most common findings in symptomatic CM1 among children aged younger than 3 years.
- Sensorimotor deficits, lower cranial nerve dysfunction, and respiratory changes in CM1 relate to syringomyelia and compression at the cervicomedullary junction.

INTRODUCTION

The definition of Chiari 1 malformation (CM1) continues to evolve. Diagnosis is often based on imaging features and include herniation of the cerebellar tonsils (one or both) 5 mm or more below a line drawn from the basion to the opisthion (McRae's line), herniations of 3 to 5 mm in the presence of syringomyelia, or if there is a peg-like rather than rounded appearance to the tonsils.[1] Tonsillar impaction occurring at the foramen magnum with consequent disruption of cerebrospinal fluid (CSF) flow dynamics and compression of structures at the cervicomedullary junction leads to signs and symptoms of CM1.[2–7] The clinical features of CM1 can differ based on the extent of these disruptions, the age at presentation, and from one individual to another. Here, we present possible clinical manifestations of CM1 in order to better recognize and care for individuals with CM1.

APPROACH

A complete history and physical examination are necessary when assessing children with CM1. History from the child should be obtained whenever possible; localization and association with certain activities can be extremely informative. The first important consideration is to distinguish *incidental* from *symptomatic CM1*. The second is to recognize the *associated features* of CM1 (eg, syringomyelia and scoliosis) and to look for *secondary causes* of tonsillar herniation (ie, acquired Chiari malformation) where different management strategies may be indicated. Secondary causes can include mass lesions,[8] craniosynostosis,[9] intracranial hypertension (eg, idiopathic intracranial hypertension),[10] intracranial hypotension (eg, CSF leak or lumboperitoneal shunting),[11,12] and less understood associations such as hydrocephalus[13] and tethered cord.[14]

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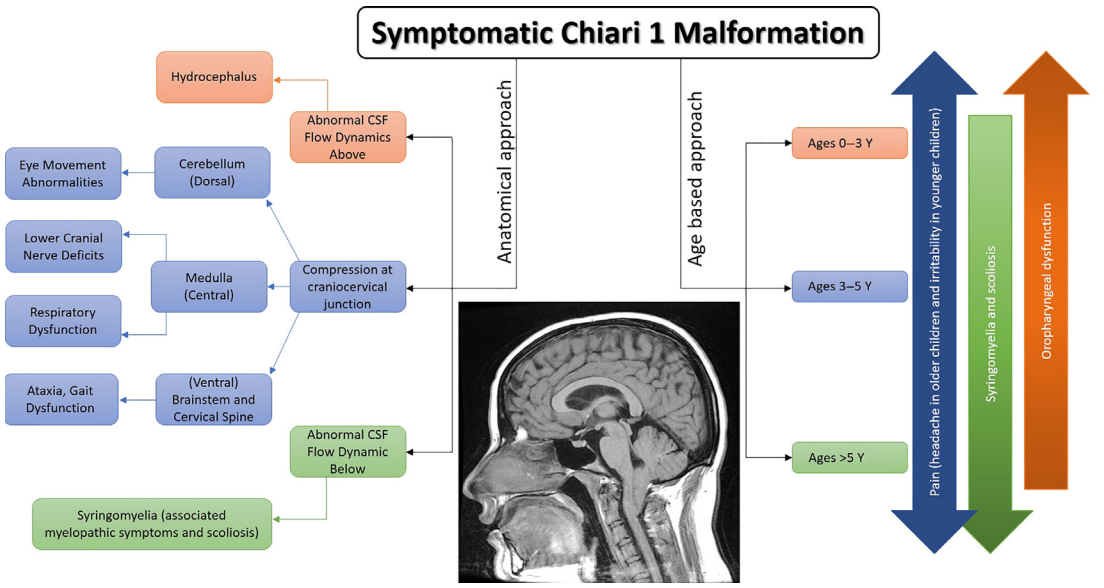

Fig. 1. Chiari 1 malformation (midsagittal MRI scan of head and upper cervical spine). Age-based and anatomic approaches to manifestations of Chiari 1 malformation. CSF, cerebrospinal fluid.

ASYMPTOMATIC CHIARI 1 MALFORMATION

An incidental CM1 may be seen in children undergoing MRI for other reasons approximately 0.2% to 1.3% of the time.[15,16] These radiological findings may improve or even resolve as the child grows, and progression of symptoms is infrequent.[15,17] Approximately 15% to 37% of pediatric patients with radiographic evidence of inferior cerebellar tonsillar ectopia may be asymptomatic, based on large retrospective reviews.[18–20] The natural clinical history of these individuals continues to be studied but mounting evidence suggests that asymptomatic children largely remain so during long-term follow-up,[21] and even those with mild features of CM1 are likely to remain stable or improve with time.[18,22–24] Therefore, recognizing the clinical features of CM1 is important for clinical decision-making.

SYMPTOMATIC CHIARI 1 MALFORMATION

Patients with CM1 usually present between the ages of 3 and 13 years.[17,18,21–26] Symptoms tend to be present for variable time points before finding a CM1.[26] In symptomatic patients with CM1, the following characteristics should be recognized[27]:

- Clinical presentation (symptoms and signs) varies with respect to age and
- Younger patients tend to present with shorter symptom duration than older patients.

Age-based and anatomic approaches are shown in **Fig. 1**.

Chiari Headaches

Headaches are one of the most common types of pain in children with a reported prevalence of 20% to 88%.[28] These are often tension type headaches and migraines and can occur in patients with CM1.[29,30] Headaches are a significant component of the clinical presentation in 27% to 81% of children with symptomatic CM1.[3–6,19,27,31] No specific criteria exists for diagnosing a "Chiari headache" and its characteristics continue to be updated in the International Headache Society Classification.[32] However, 3 aspects can aid the clinician in suspecting a headache caused by CM1[32]:

- Evidence of CM1 on imaging (cerebellar tonsillar herniation or crowding of the subarachnoid space around the cervicomedullary junction),
- Common headache characteristics described in CM1 (see later discussion), and
- Presence of associated symptoms and signs of CM1 (eg, intermittent dizziness, symptoms related to syringomyelia and/or scoliosis, symptoms related to compression of structures at the cervicomedullary junction).

Classically, headaches in adolescents and adults with CM1 demonstrate the following characteristics[2,3,5,6,18,19,33–36]:

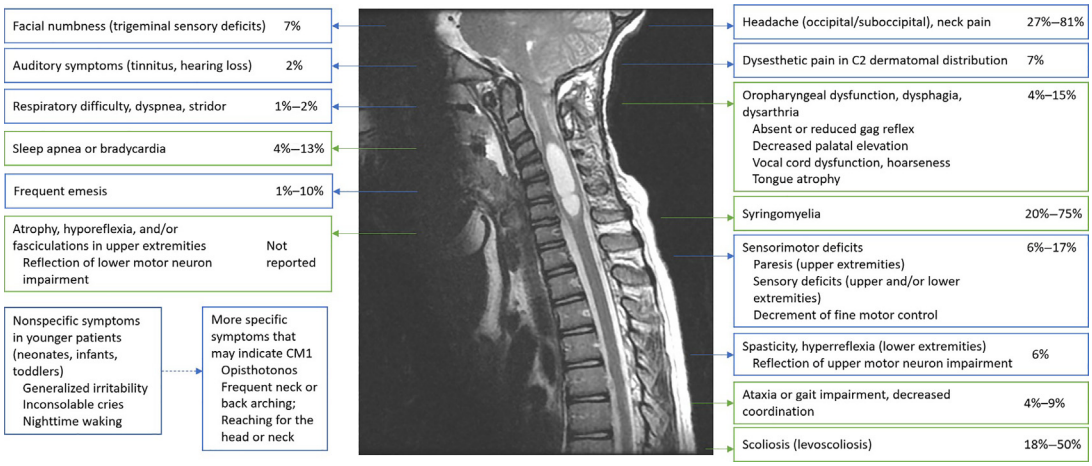

Fig. 2. Chiari 1 malformation with associated cervical syrinx. Common symptoms (blue) and signs (green) and their reported frequency (prevalence estimates are based on frequencies reported in peer-reviewed publications, and likely represent upper estimates in most cases). (*Data from* Refs.[3,4,18,19,27,31,34–36,44,57])

- Most often occipital or suboccipital,
- Provoked or intensified by Valsalva-type maneuvers,
- Last less than 5 minutes, and
- Frequent and severe.

Various functions or activities that induce a Valsalva-type response, consistent with childhood behavior, include the following[27]:

- Sneezing, coughing;
- Laughing, screaming;
- Defecation; and
- Running, repetitive jumping.

The description of the headache is variable (eg, pounding, crushing, pressure-like, pulsatile, radiating to vertex, behind the eyes, or to the neck and shoulders), and other aggravating factors reported include head movement (eg, turning, extending), change in position, and physical exertion. It is important to note that most patients with symptomatic CM1 tend to exhibit more than one symptom rather than headache in isolation (although this can occur)[26,37] and that those with the "classic" headache characteristics are more likely to respond to surgical decompression.[37] However, a retrospective study suggests that some individuals with CM1 may present with poorly localized headaches or frontotemporal headaches and still benefit from surgical decompression (although less frequently).[38] Additionally, younger patients (ie, neonates, infants, toddlers) may fail to demonstrate or adequately communicate these classic headache descriptors,[19] and therefore other symptoms and signs should be recognized as will be discussed.

Scoliosis and Syringomyelia

In addition to headaches and/or neck pain, the most common radiographic associations with CM1 are syringomyelia and scoliosis (most commonly levoscoliosis), common signs and symptoms and their frequency are shown in **Fig. 2**.[3,18,19,27,34,36] In children undergoing craniospinal MRI scans (most common indications were scoliosis and pain) and found to have a syrinx, CM1 was the most common concurrent diagnosis in 43%.[39]

Syringomyelia in CM1 demonstrates predilection for the following regions[2,3]:

- Cervical spinal cord (15%–21%),
- Cervicothoracic spinal cord (12%–25%),
- Thoracic spinal cord (15%–16%),
- Lumbar spinal cord (3%–4%), and
- Holocord (39%–44%).

Classic symptoms and physical examination signs suggesting syringomyelia include the following[5,27,35]:

- Upper extremity weakness, prominently affecting intrinsic muscles of the hand,
- Pain and temperature sensory loss (functions served by the spinal cord anterolateral spinothalamic tracts) in a "cape-like" distribution,
- Preservation of light touch sensation and proprioception (functions served by the spinal cord dorsal columns), and
- Absence of superficial abdominal reflexes ipsilateral to the convexity of scoliosis.

Characterized objectively by Cobb angles, scoliosis demonstrates a strong association with

syringomyelia in CM1.[6,7,27,34,36] Scoliosis can be identified in 31% of children with CM1 and syringomylia.[25] In general, the following rules apply[3,7,34]:

- Most (but not all) pediatric patients with CM1 with scoliosis have underlying syringomyelia and
- Not all patients with CM1 with syringomyelia have scoliosis.

These patients may report back or shoulder pain, paresthesias, gait disturbance, and/or clumsiness. Alternatively, physical examination findings of cosmetic irregularity along the spine, asymmetry of the shoulders or pelvis, subtle sensorimotor deficits, or hyperreflexia may reflect underlying scoliosis or syringomyelia.

COMPRESSION OF THE CERVICOMEDULLARY JUNCTION

Less common but concerning symptoms of CM1 include those related to compression of structures around the cervicomedullary junction[2–4,27,35,36]:

- *Medullary compression* may adversely affect respiratory function and lead to sleep apnea,
- *Brainstem or cervical spinal cord compression* can cause sensorimotor deficits (eg, hemiparesis, upper extremity paraparesis, quadriparesis), spasticity, or bladder dysfunction,
- *Lower cranial nerve deficits* (present in up to 10% of pediatric patients with CM1) can cause dysphagia, absent gag reflex, dysarthria, vocal cord dysfunction, or abnormal extraocular motility (eg, esotropia owing to cranial nerve VI paresis)[2,3,5,27,36];
- *Cerebellar flocculus compression* may present as gaze-evoked nystagmus in up to 30% of young adults with CM1,[36] or less commonly other neuro-ophthalmological findings,[40] and its incidence in the pediatric population is not as well defined but reported[41], and
- Up to 10% of those with CM1 may also present with *hydrocephalus* owing to fourth ventricular outlet obstruction.[3,27]

These symptoms occur with less frequency in the modern era likely owing to the advent of MRI offering earlier diagnosis, before neurologic disability occurs.[2,27] However, lower cranial nerve dysfunction is a predominant presentation in children aged younger than 3 years with symptomatic CM1.[36] Medullary compression symptoms are also seen more commonly in children with complex CM1 that includes basilar invagination and/or instability with ventral compression of the cervicomedullary junction.[2,42]

CLINICAL PRESENTATION DURING THE NEONATAL PERIOD AND INFANCY (AGE 0–3 YEARS)

The symptoms and signs of CM1 in this population are more commonly related to pain and features of compression at the craniocervical junction. Pain can present as generalized irritability, inconsolable cries, and nighttime waking in most patients.[3–5,18,27,43] Additional clinical signs can provide further localization, and include the following[3–5,18,27,36]:

- Opisthotonos,
- Frequent neck or back extension/arching, and
- Crying spells with behavioral patterns suggesting head discomfort (eg, reaching for the head or neck).

Oropharyngeal dysfunction, caused by medullary compression and lower cranial nerve compromise, is one of the most common presenting symptoms in approximately 62% to 78% of children aged 0 to 2 years with CM1.[34,36,43] Clinical manifestations can include the following[4,5,20,27,34,36,42]:

- Dysphagia, choking, or aspiration;
- Poor feeding, failure to thrive;
- Gastroesophageal reflux;
- Persistent cough;
- Snoring or episodic sleep apnea;
- Stridor; and
- Recurrent respiratory infections.

Less common symptoms include abdominal pain and vomiting.[36] These more dramatic presentations are typically observed in the setting of significant ventral compression owing to basilar invagination, retroflexion of the dens, and/or frank cervical instability.[42,44] Diagnosis of CM1 in this age group is low and likely reflects delayed identification among other factors. It is not uncommon for these children to undergo gastrointestinal investigations and interventions (eg, feeding tubes), or assessments for vocal cord paralysis that require tracheostomies before the identification of CM1.[36,45]

Clinical presentations with syringomyelia and/or scoliosis are possible in the neonatal and infant populations but occur less frequently than in older children.[4,34,36] In this and other age groups, there does not seem to exist a correlation between the extent of tonsillar herniation and the presence or absence of

syringomyelia.[34] Long tract signs can also be present in this age group such as spasticity and gait instability.[43]

CLINICAL PRESENTATION IN TODDLERS (AGE 3–5 YEARS)

As patients develop improved ability to communicate and localize their pain, they may be able to more effectively verbalize complaints attributable to the presence of an underlying CM1. Toddlers with adequate verbal skills may report headache pain or discomfort in the upper neck. Occipital headaches represent a component of the clinical presentation of CM1 in up to 40% to 78% of patients during the toddler stages and early childhood.[3,4,34,36,46] Children aged 3 to 5 years with CM1 frequently present with syringomyelia and/or scoliosis (the latter present in 14%–38% of pediatric patients).[4,27,31,34,36] As with neonates and infants, symptoms or signs of medullary and lower cranial nerve dysfunction (eg, sleep apnea, oropharyngeal dysphagia, dysarthria, absent gag reflex) may be present in toddlers with CM1.[19,27,34,36]

CLINICAL PRESENTATION DURING CHILDHOOD AND ADOLESCENCE (AGE 5 YEARS AND OLDER)

As they mature into the childhood and adolescent years, pediatric patients more frequently and reliably report the classic symptoms of CM1. Most commonly, patients note occipital headaches and/or neck pain, often induced by Valsalva-type maneuvers (eg, straining for a bowel movement, laughing, coughing, sneezing) and of short duration (typically <5 minutes).[3] To warrant surgical consideration, these headaches should be severe and frequent enough to impact activities of daily living (eg, missing school) or quality of life.[5] Some children present with frontotemporal or poorly localized headaches and still benefit from decompression but with lower success rates reported.[38] As with younger patients, older children and adolescents with CM1 may also experience oropharyngeal dysfunction, although less frequently.

Scoliosis, typically associated with syringomyelia (present in 19%–76% of patients with CM1), represents another important component of the clinical presentation in this group of patients.[2–4,7,31,34,36] Syringomyelia with scoliosis may lead to back or shoulder pain.[3] Children may also present with myelopathic features (shown in **Fig. 3**) in the absence of any headache.[38] The constellation of CM1, syringomyelia, and/or scoliosis may produce multiple physical examination findings that are easier to elicit or observe within older children and adolescents (**Fig. 4**).

CLINICAL PRESENTATION IN ADULTS

In adults with CM1, symptoms in order of frequency include headaches (cough-induced, migrainous, or others), paresthesia, ataxia, and less frequently oropharyngeal and cranial nerve dysfunction. Available, although, limited studies suggest that asymptomatic and mildly symptomatic adults with CM1 have a benign natural history. Indications for intervention vary but include Chiari headaches affecting the quality of life, neurologic symptoms attributed to brainstem compression, and symptomatic or enlarging syrinx.[47]

CLINICAL CONDITIONS ASSOCIATED WITH CHIARI I MALFORMATION

In addition to scoliosis, other, less common spinal or craniocervical osseous abnormalities may occur in the pediatric CM1 population (**Table 1**).[2–6,27,31,34,42,44,48] CM1 represents a common finding (up to 33%–38%) in patients with craniovertebral junction abnormalities such as basilar invagination.[2] A variety of other clinical conditions and syndromes have been associated with CM1, many of which may be incidental or displacement of the tonsils occurs secondary to the primary disease.[49] Common associated conditions include the following[3–5,27,34]:

- Hydrocephalus (8%–10%),[3,27]
- Craniosynostosis,[50]
- Neurofibromatosis type I (up to 5%),[3,27]
- Ehlers Danlos syndrome type 3,[51,52] and
- Growth hormone deficiency (idiopathic; around 4%).[3,27]

RARE AND ACUTE PRESENTATIONS OF CHIARI I MALFORMATION IN CHILDREN

In addition to the common clinical presentations of CM1 described, more obscure presentations exist based on their acuity, rapid progression, or rarity of symptomatology. In unusual circumstances, pediatric patients with CM1 can present acutely in distress and require urgent operative intervention. Previously reported acute onset or rapidly progressive symptoms and signs include the following[3,53–56]:

- Dysphagia,
- Isolated upper or upper and lower extremity weakness or paralysis,
- Paresthesias,

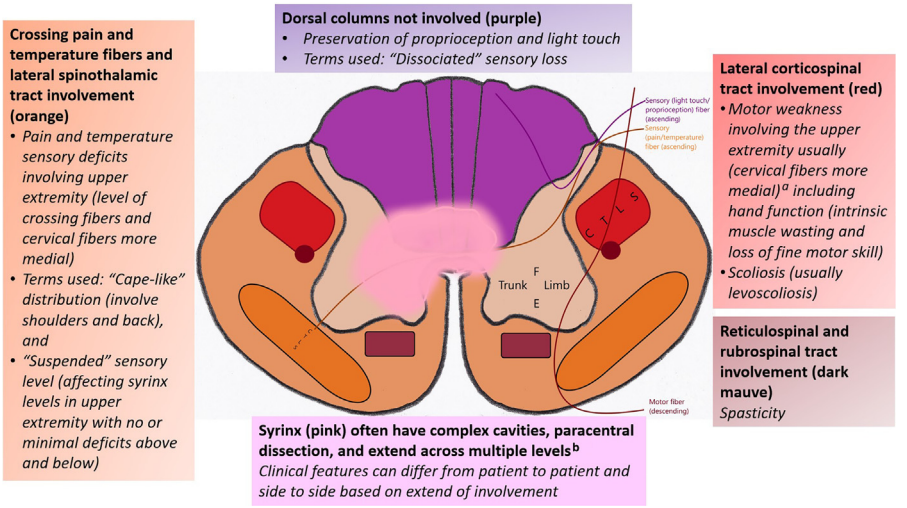

Fig. 3. Clinical features associated with syringomyelia. Authors' depiction of cervical spinal cord cross section with select tracts represented. [a]This anatomic pathophysiology has recently been challenged.[58] [b]Pathological features of syrinx from Ref.[59] C, cervical; E, extensors; F, flexors; L, lumbar; S, sacral; T, thoracic.

- Respiratory distress,
- Gait dysfunction,
- Foot drop, and
- Anisocoria.

These presentations tend to have the following commonality in addition to CM1[56]:

- Often (but not always) have an inciting event (eg, trauma, infection, breath holding),
- Often (but not always) associated with spinal cord changes (syringomyelia or "presyrinx") in cases of sensorimotor deficits,
- Most respond well to surgical decompression, and
- These events are rare.

These acutely presenting or rapidly progressive symptoms and signs reflect pathologic compression of the brainstem and/or spinal cord long tracts.[53] Although the specific symptoms may not represent rare findings given their appearance as chronic symptoms in other patients with CM1, their rapid presentation or progression defy common patterns. Several rare presentations of CM1 have been reported in the literature and are summarized in **Table 2**.[3–5,19,20,27,31,34–36,53]

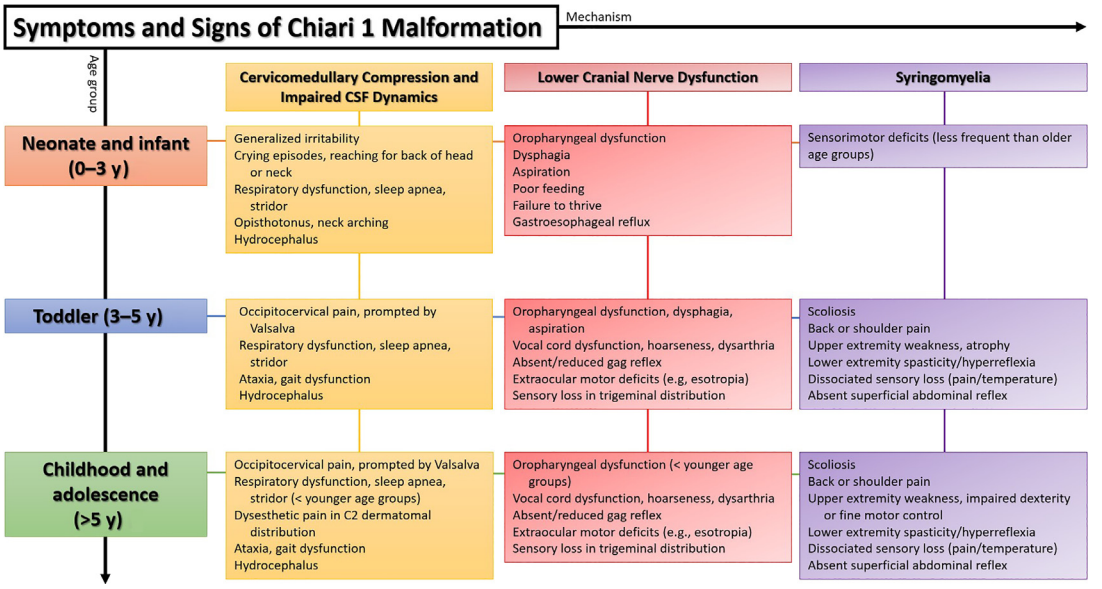

Fig. 4. Prominent symptoms and signs of Chiari I malformation and syringomyelia in children, based on mechanism and age. CSF, cerebrospinal fluid; Yrs, years.

Table 1
Abnormalities associated and/or reported with Chiari I malformation (list not exhaustive)[3,4,8–14,60]

Type/Location	Association
Cranial vault	Craniosynostosis Decreased volume of posterior fossa
Intracranial	Hydrocephalus Idiopathic intracranial hypertension Mass lesions
Skull base	Fibrous dysplasia and McCune-Alright syndrome Achondroplasia
Craniocervical junction	Basilar invagination Platybasia Atlas assimilation
Cervical spine	Odontoid process retroversion/ retroflexion Klippel–Feil anomaly or variants
Vertebral column/ spine	Hemivertebra Butterfly vertebra Tethered cord
Skeletal	Cervical rib Sprengel deformity
Syndromes	Neurofibromatosis type 1 Ehler's Danlos syndrome
Others	Idiopathic growth hormone deficiency Epilepsy

Table 2
Rare presentations of Chiari I malformation in children

Severity of Presentation	Clinical Finding
Mild	Nystagmus (typically down-beating) Chronic hiccups Chronic cough Cerebellar or cerebellovestibular dysfunction (eg, vertigo)
Moderate	Focal sensorimotor deficits (mononeuropathy; eg, plantar flexion weakness) Urinary incontinence Torticollis Trigeminal or glossopharyngeal neuralgia Sensorineural hearing loss
Severe	Syncopal episodes, drop attacks Acute spinal cord injury after trauma (eg, quadriplegia) Respiratory failure requiring mechanical ventilation Cardiorespiratory arrest, sudden death

SUMMARY

Incidental CM1 is common and likely to remain asymptomatic in most cases. When symptomatic, it most often presents with headaches that are typically occipital or suboccipital, provoked or worsened by Valsalva-like maneuvers, and of relatively short duration. CM1 is most commonly associated with syringomyelia with or without scoliosis at presentation in the pediatric population. CM1 and syringomyelia may be associated with a wide spectrum of signs and symptoms in children, which can differ in presentation between older and the very young children. In those younger than 3 years, in addition to pain symptoms, oropharyngeal symptoms seem to predominate more so than syringomyelia or scoliosis. Other symptoms related to compression at the cervicomedullary junction include respiratory dysfunction, lower cranial nerve deficits, ataxia, and nystagmus. CM1 is also associated with osseous abnormalities and certain genetic syndromes. Although most patients with CM1 with symptoms tend to have a longer duration of symptoms and benign course, acute neurologic changes have been reported and are rare.

CLINICS CARE POINTS

- Evaluation of children referred for incidental or asymptomatic CM1 should rule out associated findings and management should consider that most children will remain asymptomatic over time.

- The nature of the headache in symptomatic CM1, its effect on daily activities, and other signs and symptoms related to CM1 should be elicited on history and examination to identify patients that are likely to benefit from surgical decompression.
- There should be a high clinical suspicion for cervicomedullary compression, including CM1, in patients younger than 3 years presenting with pain symptoms, oropharyngeal dysfunction, and signs such as opisthotonos, neck/back arching, or reaching for the head or neck.
- Syringomyelia and scoliosis are highly associated with CM1 and signs and symptoms (eg, sensorimotor deficits, shoulder or back pain, asymmetry of the shoulder, chest, or pelvis) should be elicited during assessment.

DISCLOSURE

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