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Seminars in
Pediatric
Neurology

Headache and Chiari I Malformation in Children and Adolescents

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Headache is a common problem in children and adolescents. Its recurrent and disabling nature may lead to use of neuroimaging to exclude secondary causes of headache such as Chiari I malformation (CM I). CM I has a variety of presentation with headache being the most common symptom. CM I can be asymptomatic and is also often found incidentally in neuroimaging done for conditions other than headache. This article reviews the spectrum of headache in patients with CM I.
Semin Pediatr Neurol 23:35-39

Introduction

Chiari I malformation (CM I) is a condition characterized by downward displacement of the cerebellar tonsils of at least 3-5 mm through the foramen magnum into the spinal canal. In the general population, its prevalence is 0.6%-0.9 %; and ~1% in children.[1-3] It has a variable clinical presentation with headache being the most common initial symptom. The headache in CM I is poorly defined in the literature especially in the pediatric population.

Headache, in general, is a common symptom in children and adolescents that leads to neurologic consultation. Its recurrent and disabling nature raises anxiety among families and concern for secondary cause of headache leading to wide use of neuroimaging in the evaluation of these patients. The rate of brain abnormalities in pediatric patients with headache is 14%-28%. Most abnormal findings do not change the headache management.[4,5] CM I is identified in about 5.8% of patients imaged for headache.[5] The coexistence of a primary headache disorder and CM I, or whether CM I is causative of the headache is a challenge to many medical providers.[5]

Signs and Symptoms

Symptoms from CM I usually start in the second or third decade of life, but there is increased recognition of symptomatic cases in children. Greenlee et al[6] reported the spectrum of presentations of CM I in children less than 6 years. The astounding observation is the presence of abnormal oropharyngeal function manifesting as cough, stridor, dysphagia, abnormal vocal cord movement, gastroesophageal reflux, aspiration, recurrent respiratory infection, feeding intolerance, and failure to thrive among children less than 3 years of age. Symptomatic CM I patients more than 3 years present with scoliosis or headache worsened by Valsalva maneuver.[6] Beyond this age group, the clinical manifestations of CM I in pediatric and adult patients appear similar. The most common presenting symptom is headache. Other manifestations are neck pain, vertigo, sensory changes, ataxia, syncope, sleep apnea, and drop attacks.[7-11]

CM I has 3 types of neurologic signs: (1) brainstem syndrome such as respiratory irregularities, nystagmus, lower cranial nerve dysfunction, tongue atrophy, and facial sensory loss; (2) spinal cord dysfunction as a result of direct cord compression or a syrinx. This can include motor and sensory losses especially in the hands, hyperreflexia or hyporeflexia and urinary incontinence; and (3) cerebellar syndrome such as truncal and appendicular ataxia. Progressive scoliosis is also found to be a common CM I presentation in the presence of syringomyelia.[8,10]

Radiologic Findings

The widely accepted radiographic definition is (1) herniation of 1 or both tonsils >5 mm below the foramen magnum; (2) bilateral tonsillar herniation of 3-5 mm accompanied by features of subarachnoid space crowding in the craniocervical junction such as compression of the cerebrospinal fluid (CSF) spaces posterior and lateral to cerebellum, reduced height of the supraocciput, increased slope of the tentorium

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1071-9091/16/$-see front matter
http://dx.doi.org/10.1016/j.spen.2016.01.001

Table 1 ICHD-III (Beta Version) Criteria for Headache Attributed to CM I

A. Headache fulfilling criterion C
B. Chiari I malformation type I (CM I) has been demonstrated
C. Evidence of causation demonstrated by at least 2 of the following:
1. Either or both of the following:
a. Headache has developed in temporal relation to the CM I
b. Headache has resolved within 3 mo after successful treatment of the CM I.
2. Headache has at least 1 of the following 3 characteristics:
a. precipitated by cough or other Valsalva-like maneuver
b. Occipital or suboccipital location
c. Lasting <5 min.
3. Headache is associated with other symptoms and clinical signs of brainstem, cerebellar, lower cranial nerve, and cervical spinal cord dysfunction.
D. Not better accounted for by another ICHD-III diagnosis.

and kinking of the medulla oblongata.[8,11] A significant proportion of patients have syringomyelia, and it has been seen in 12% of children at the time of initial CM I diagnosis.[7]

Headache and CM I

Headache is the most common symptom of CM I with frequency in adults ranging from 30%-80%[3,10,11]and 15%-75% in children.[2,7,12-14] The headache phenotype in CM I is ill-defined in the literature especially in children, and the information is mostly based on retrospective studies. Current literature shows that there is a spectrum of headache associated with CM I.

Headache Attributed to CM I

Headache caused by CM I is classically described as occipital or suboccipital in location, brief in duration and triggered by cough, Valsalva maneuver, or physical activity.[2,3,10,11,15] Although headaches provoked by cough or Valsalva maneuver can be a primary headache disorder, it is a caveat that secondary causes should first be ruled out. The studies of Pascual et al[15,16] showed that all symptomatic cases of cough headache were associated with CM I; and most of them were associated with other posterior fossa signs and symptoms. The headache duration was variable, from seconds to several weeks. Chen et al[17] also reported posterior fossa lesions as the most common etiology for secondary cough headache. As such, headache caused by CM I is often descriptively akin to primary cough headache except that it may possibly be longer in duration (minutes rather than seconds).[12,18]

Toldo et al[19] classified headache among children with CM I using the international classification of headache disorders-II (ICHD-II) criteria. Among the 45 patients with CM I, only 27% of them had headache attributed to CM I (CMH). Most cases had tonsillar ectopia more than 9 mm. There were no cases of CMH among those with borderline tonsillar ectopia. The study also found relevant associations between the extent of tonsillar ectopia and CMH clinical characteristics, such as brief duration of pain, occipital in location, and headache triggered by physical activity and Valsalva maneuver. Only 3 patients were surgically treated due to headache and worsening of other signs and symptoms of CM I. After surgery, 2 had complete resolution of headache and 1 had reduction of daily headache. To date, this study best defined the headache pattern in children with CM I and proposed that the presence of CMH along with 3 other signs or symptoms of CM I were highly predictive of severe tonsillar ectopia.[3] Table 1 lists the ICHD third edition beta version (ICHD-III beta) criteria for the diagnosis of CMH.[20]

The pathophysiology of headache from CM I remains unclear but several mechanisms underlying headache associated with cough in patients with CM I have been proposed. Pascual et al[15] attributed the headache to the compression of C1 and C2 roots by further tonsillar herniation occurring during Valsalva-like maneuvers. It has also been thought to be a result of craniospinal pressure dissociation or due to sudden increased intrathecal pressure caused by obstruction to the free flow of CSF in the subarachnoid space.

Primary Headache Disorder Coexisting With CM I

Headache is a common complaint among children and adolescents. Its prevalence increases throughout childhood and up to 75% of children have reported significant headache by 15 years of age.[21,22] The overall prevalence of pediatric migraine is 7%; about 10% for girls and about 5% for boys. The prevalence of tension type headache (TTH) is 0.9%-24%.[23,24] Chronic daily headache (CDH) has now become a well-recognized primary headache disorder in the pediatric population and has a prevalence of 3.5%.[25,26] As such, there is a high likelihood of primary headache disorder coexisting with other conditions such as CM I. In addition, the ease in obtaining brain magnetic resonance imaging (MRI) has led to the incidental finding of CM I among headache patients making clinical decision challenging.