

Chiari Malformation Presenting as Headaches Associated with Laughter

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Abstract

Presentation

We describe the case of a 5 year old boy, referred to our outpatient department with a one year history of headaches associated with laughter.

Diagnosis

Investigation with MRI Brain revealed Chiari Type 1 Malformation (CM-1), with cerebellar tonsillar descent of 19mm below the foramen magnum.

Treatment

He is being managed conservatively with serial neuroimaging and symptom monitoring.

Discussion

CM-1 is a hindbrain malformation characterised by ≥ 5 mm herniation of the cerebellar tonsils.^{1,2} It is diagnosed radiographically, and is increasingly being detected incidentally.^{3,4} The natural history of asymptomatic patients is usually to remain asymptomatic, and symptomatic patients often show symptom improvement, particularly in paediatric populations.^{3,5} Neurosurgical interventions may be offered based on symptoms and radiographic findings, but carries a complication rate of 8.2%.⁶

Introduction

Chiari malformations describe a range of hindbrain malformations characterised by downward herniation of the cerebellar tonsils.¹ CM-1 is defined as at least 5 mm of tonsillar herniation below the foramen magnum.²

Confirmed CM-1 is commonly associated with other conditions such as syringomyelia (23.8%-65%), scoliosis (11.5%-42%), basilar invagination (12%), hydrocephalus (5.9%) and tethered cord syndrome (2.2%).^{1,7}

The true prevalence of CM-1 remains unknown, as an unidentified proportion of individuals remain asymptomatic and undiagnosed. Increasingly, CM-1 is diagnosed incidentally - a recent study showed 37% of children diagnosed with CM-1 had neuroimaging for symptoms unlikely to be related to CM-1.^{2,3,4}

In symptomatic individuals, the mean age of symptom onset is 24.9 years, with SD of 15.8 years.¹ 31% of patients are diagnosed in paediatric services, aged <18 years.⁵ Amongst paediatric patients, older age at diagnosis predicted worse neurological symptoms.²

Young adults with CM-1 classically present with occipital pain precipitated by cough or Valsalva manoeuvres.² Paediatric patients may also present with headache, however cough headaches and occipital location are less common (4% and 6% respectively).² The most common presenting symptoms in children are headaches (55%), neck pain (12%), vertigo (8%), sensory changes (6%) and ataxia or poor coordination (6%).²

Case Report

This 5 year old boy experiences sudden onset generalised headaches of variable severity, associated predominantly with laughter, and once with shouting. They have not occurred with coughing, sneezing, straining or bending over. The pain is described as a tightening sensation around his entire head, which may be associated with pallor, nausea, gagging, or generalised weakness. His reaction to the pain is to grasp his head with both hands and sit or lie on the floor. The pain lasts for several seconds then subsides, with return to good form afterwards. These headaches have not interfered with schooling or hobbies, including soccer and GAA. At the time of initial assessment, they were occurring daily.

Neurological examination was unremarkable.

MRI Brain revealed Chiari Type 1 Malformation, with cerebellar tonsillar herniation 19 mm below the foramen magnum (see image 1). This was an isolated finding on imaging, with no other abnormalities noted.



Image 1: MRI Brain showed Chiari Type 1 malformation with tonsillar descent 19 mm below to foramen magnum.

A referral was made to a tertiary neurological centre for further management. At the time of their review, his headaches had reduced in frequency although were increasing in severity. His parents are currently keeping a diary of headache frequency, severity and precipitants. He will be followed up by the Neurosurgical team with serial MRI brain and whole spine and regular review of symptoms.

Discussion

Laughter is a rare precipitating factor for headaches.⁸ Although case reports exist of laugh headaches as the presenting feature of CM-1 in adults, to our knowledge this is the first published paediatric case report. There is currently no published data specifically documenting the frequency of isolated laugh headaches as the presenting symptom of CM-1.

The natural history of asymptomatic CM-1 is that most patients remain asymptomatic (93.3%).³ In symptomatic CM-1 managed conservatively, the natural history is improvement in headaches and nausea for many patients (37-40% and 89% respectively), with paediatric patients more likely to show improvement than adults (67-71% headache improvement).^{3,5} Patients experiencing ataxia and sensory disturbances are less likely to improve without surgical interventions.^{3,5}

The decision to offer neurosurgical intervention is based on symptoms and neuroimaging.⁴ Controversy exists regarding the precise indications for surgery, timing and type of interventions offered, and clinical and radiographic outcomes.^{2,6} Neurosurgical management carries a complication rate of 8.2%, most commonly nervous system related (2.8%), anaemia (2.4%) or acute respiratory distress (2.1%).⁶

In light of the natural history of CM-1 and not insignificant complication rate with neurosurgical interventions, it is reasonable to manage asymptomatic and mildly symptomatic CM-1 patients conservatively with serial imaging and review of symptoms.³

Patient Consent: Received

Declaration of Conflicts of Interest:

The authors have no conflicts of interest to declare.

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