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Short Communication

Interdisciplinary Approach of Patients with Chiari type I Malformation and Headache - Experience at Hospital de Clínicas José de San Martín

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Abstract

Chiari type I malformation (CM1) is characterized by the caudal displacement of the cerebellar tonsils in a cone-shaped projection through the foramen magnum into the cervical spinal canal. Headache is the most common and at times only symptom. In a patient with headaches, neuroimages may be necessary to rule out a secondary cause of the pain. If CM1 is found on imaging studies, headache is generally attributed to the malformation and the patient is referred to the neurosurgeon for surgical intervention. Although headache is the most common symptom of CM1, the malformation is often an incidental finding in a pain study and there is no causal relation between both entities. Therefore, in an interdisciplinary approach, the Departments of Neurology and Neurosurgery evaluated those patients with headache and evidence of CM1 on brain magnetic resonance imaging (MRI), as part of the surgical work-up to characterize the type of pain and thereby to determine the most appropriate therapeutic approach. The results showed that in 85% of the patient's headache was not secondary to CM1, and surgical resolution was no longer an option in these cases.

ABBREVIATIONS

CM1: Chiari type I malformation; MRI: Magnetic resonance imaging; IHS: International Headache Society; CSF: Cerebrospinal fluid; HII: Idiopathic intracranial hypertension. ICHD-3: The International Classification of Headache Disorders 3rd Edition

INTRODUCTION

Chiari malformation was first described in 1891 by the Austrian pathologist Hans Chiari who defined the entity as a group of syndromes consisting of different diseases of posterior fossa development [1].

Currently, Chiari malformations are classified according to the degree of herniation and their association with neural defects [2]. Traditionally, four types were identified (I-IV) and types 0, 1.5, and 5 were later added [3,4] (Table 1). Type I (CM1) is the most common Chiari malformation. CM1 is generally congenital caused by structural defects secondary to a genetic mutation, a vitamin deficit, or a nutritional deficit in the maternal diet [1].

CM1 is characterized by the descent of the cerebellar tonsils in a cone-shaped projection into the cervical spinal canal through the foramen magnum without displacement of the brain stem. In adults, displacement of the cerebellar tonsils into the spinal canal of 3 mm or less is considered normal, while a descent of 5 mm or more is considered pathological. A descent between 3 and 5 mm is considered "borderline" and should be judged as significant if there is evidence of occupation of the subarachnoid space in the area of the craniocervical junction shown by the presence of compression of the spaces posterior and lateral to the cerebellum, reduced height of the supraocciput, increased slope of the tentorium, or compression of the medulla oblongata [5-9].

A data base of magnetic resonance imaging studies revealed a descent of the cerebellar tonsils of at least 5 mm in 0.7% of the population [5]. The clinical context of CM1 is relevant, as many of the individuals are asymptomatic[8] and it is often an incidental finding on imaging studies. Only about 0.01-0.04% of adults demostrate symptoms and MRI evidence of CM1 [6]. Epidemiologic research on adult patients with Chiari type 1 showed a 3:1 female to make preference for the disease with a higher prevalence of CM1 in Pacific Islanders [5].

Although neck pain (12%), vertigo (8%), and ataxia (6%) [5] are also common symptoms; headache is far more frequent in the symptomatic patient (15% to 98%) [2]. Headache is characterized by a localized pain at the suboccipital-occipital level that is triggered by a Valsalva maneuver [7], is short-lasting (less than 5 minutes) and resolves after adequate treatment of the Chiari malformation [9]. The International Headache Society

(IHS) has established the diagnostic criteria to define this type of headache [9] (Table 2).

Other primary or secondary headaches may be incidentally found in patients with imaging findings of CM1. Thus, in an interdisciplinary approach the Departments of Neurology and Neurosurgery evaluated patients with headache and CM1 to determine if the pain was secondary to the structural defect or if it was due to another cause and based on these data define the most appropriate therapeutic option.

MATERIALS AND METHODS

In the Headache Unit of the Department of Neurology, 113 patients with imaging diagnosis of CM1 and headache were studied referred through the Program for Craniospinal Joint Diseases from the Department of Neurosurgery between March 2017 and June 2020. All patients underwent MRI of the cervicodorsal spine to rule out syringomyelia and to assess the dynamics of cerebrospinal fluid (CSF) flow.

CM1 was defined as the caudal descent of more than 5 mm of the cerebellar tonsils below the foramen magnum on brain MRI in a sagittal view (Figure 2a). A descent between 3 and 5 mm (borderline) was considered to be significant if it was associated with any sign of occupation of the posterolateral CSF spaces of the cerebellum, reduced height of the supraocciput, increased slope of the tentorium, or compression of the medulla oblongata [10]. Patients with a history of surgery for CM1 and those with syringomyelia were excluded from the study.

Of 113 patients with imaging findings of CM1 and headache, 22 were excluded because of a previous surgery for CM1 (15 patients) and/or symptoms of syringomyelia (7 patients). A total of 91 patients were included in the study, with a clear female predominance (F/M male ratio: 73/18) and ages between 17 and 70 years.

Through a semistructered interview and a complete physical and neurological exam, different types of headache were identified according to the criteria of the IHS (ICHD third edition) [9]. A headache diary was handed out to each patient to record the episodes for the follow-up and evaluation of the outcome.

All the patients had given their written informed consent for using their data for scientific purposes.

RESULTS AND DISCUSSION

Of 91 patients who met the inclusion criteria, 85% (77 patients) had headaches unrelated to the CM1. In these cases, headache was most often secondary to analgesic abuse (n=37) followed by migraine without aura (n=26), tension-type headache (n=6), migraine with aura (n=5), dysfunction of the

Table 1: Chiari malformations: Classification.

Type	Description
0	**Syringomyelia without cerebellar tonsil descent. Both syringomyelia and symptoms resolve after craniocervical decompression.**
I	Descent of 5 mm or more of the cerebellar tonsils through the foramen magnum into the spinal canal without brain stem displacement.
1.5	**Descent of the cerebellar tonsils with similar features as in CM1 associated with brain stem displacement.**
II	Displacement of the lower part of the vermis and elongation of the IV ventricle. Generally associated with spina bifida.
III	Herniation of the cerebellum within the spinal canal. May be associated with occipital encephalocele.
IV	Cerebellar hypoplasia
5	**Herniation of the occipital lobe through the foramen magnum.**

Abbreviations: CM1: Chiari type I malformation

Table 2: Diagnostic criteria of headache attributed to a Chiari type I malformation (International Headache Society, The international classification of headache disorders, 3rd edition, 2018).

Diagnostic criteria 7.7 Headache attributed to CM1
A. Headache fulfilling criterion C
B. CM1 has been demonstrated (1)
C. Evidence of causation demonstrated by at least two of the following: 1-- Either or both of the following: **a) headache has developed in temporal relation to the CM1 or led to its discovery** **b) headache has resolved within 3 months after successful treatment of the CM1** 2- **- Headache has at least one or more of the following three characteristics:** ♌☽ **Precipitated by cough or other Valsalva--like maneuver** ♍☽ **Occipital or suboccipital location** ♎☽ **Lasting <5 minutes** 3- **- Headache is associated with other symptoms and/or clinical signs of brainstem, cerebellar, lower cranial nerve and/or cervical spinal cord dysfunction (2)**
D. Not better accounted for by another ICHD--3 diagnosis (3)
Abbreviations: CM1: Chiari type I malformation; ICHD-3: The International Classification of Headache Disorders 3rd Edition

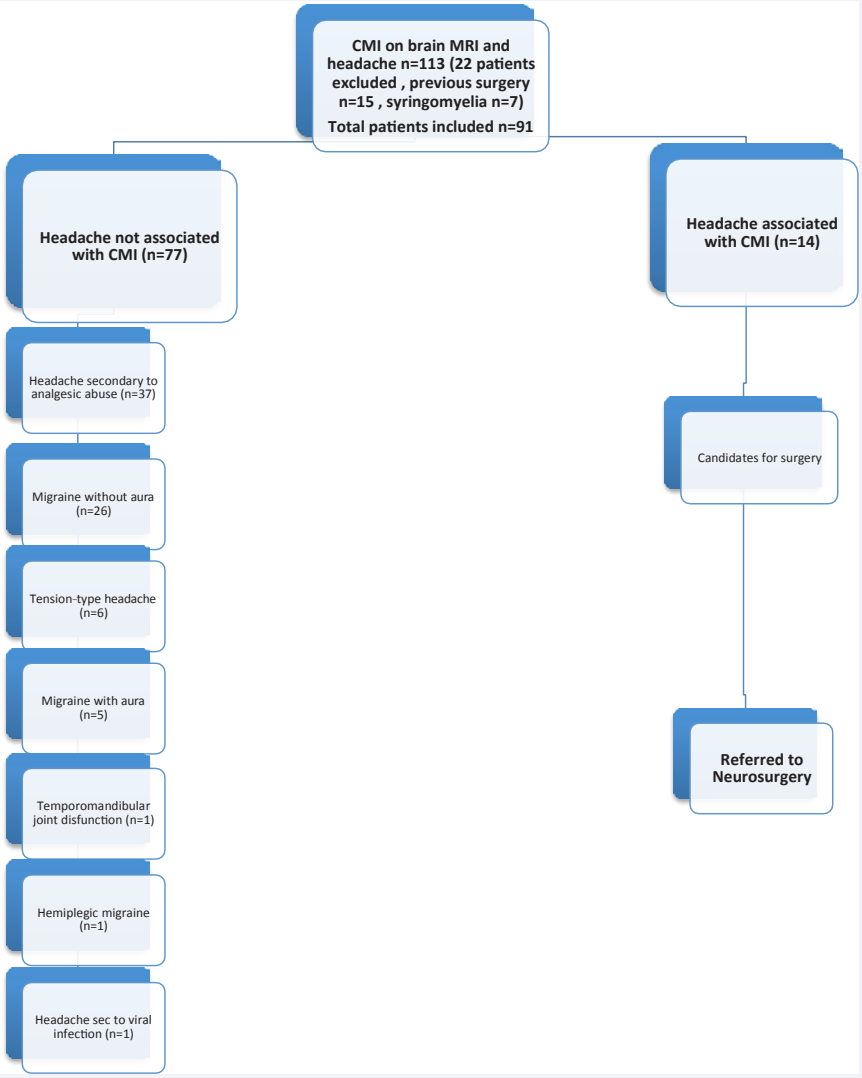

Figure 1 Results.

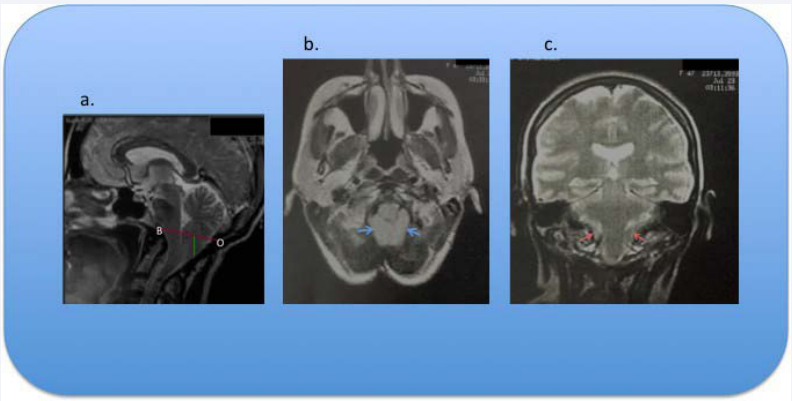

Figure 2 Brain MRI: T2 weighted images, sagittal (a), axial (b), and coronal (c) planes.
Figure (a) shows a line (purple) from the basion (B) to the opistion (O) indicating the lower limit of the posterior fossa, the reference point for measuring cerebellar tonsil ectopia. The latter is measured in mm by tracing a downwards perpendicular line (green) from the center of the cerebellar tonsils.
Figure (b) shows the position of the cerebellar tonsils in the foramen magnum (blue arrows). In the coronal view (c) asymmetrical tonsil position is observed (red arrows).

temporomandibular joint (n=1), hemiplegic migraine (n=1) and acute headache attributed to systemic viral infection (n=1)

Only fourteen of 91 patients (15%) met the criteria for headache secondary to CM1. Of these patients, eleven underwent surgery with a successful outcome and significant improvement of the headache, while three patients were lost to follow-up at both Departments (Figure 1).

The remaining patients were treated pharmacologically according to the type of headache with good outcomes.

The characteristics of the headaches are summarized in Table 3.

The clinical presentation of patients with Chiari I includes different neurological symptoms related to the compression of the cranial nerves or the brainstem. Nevertheless, headache is the most common and sometimes only symptom.

In this study, we found that in the majority of patients with headache and neuroimages compatible with CM1 the pain was due to a different cause. In only 15% of the patients headache was triggered by Valsalva maneuvers and met the criteria for CM1 of the IHS classification [9].

A study evaluating CSF hydrodynamics showed that occipital headache was associated with obstruction of the CSF flow at the level of the foramen magnum [10]. This finding is relevant to consider when evaluating surgical treatment when headache is the only symptom of CM1[10]. In our study, in all eleven patients who had headache related to CM1 the pain was occipital. Stovner has proposed the mechanism of the Valsalva maneuver-induced headache in patients with CM1 secondary to the transient pressure dissociation displacing the cerebellar tonsils downward through the foramen magnum, causing headache [11].

The development of pressure dissociation is explained by an initial elevation of spinal pressure during coughing, displacing CSF to the head. However, immediately after coughing, this displaced CSF returns with relative ease to the spinal canal in a healthy patient but not in a patient with CM1 because of impaction of the cerebellar tonsils, thereby creating a pressure dissociation between the head and spine, where intracraneal pressure is higher than that of the spine, causing pain because of traction and pressure on pain sensitive structures [12].

As previously described, CM1-related pain is short lasting (Table 2). Therefore, when continuous headache is observed in a patient with cerebellar tonsil descent on MRI, two differential diagnoses should be considered: the first possibility is headache due to intracranial hypertension secondary to hydrocephalus, which is a complication of CM1. The second is headache due to intracranial hypotension, which may cause a secondary descent of the cerebellar tonsils leading to a so-called “pseudo-Chiari”. The latter entity should be suspected in a patient with a history of lumbar puncture or recent epidural anesthesia; however, it may also occur in the context of a strenuous effort (for example, during coitus) [13]. Other conditions such as space-occupying lesions of the brainstem, cerebellum or spine and degenerative myelopathy should be considered as differential diagnosis [14].

When these two diagnoses are ruled out, the patient with continuous headache should be meticulously evaluated taking into account the IHS Criteria [9] to rule out other secondary (headache secondary to analgesic abuse) or primary causes (migraine, tension-type headache). Nevertheless, there is not enough evidence suggesting that CM1 could induce primary headache. First, the number of reported cases with a combination of tension-type headache or migraine and headache due to CM1 is very low [11,15]. Second, in a series of patients with symptomatic CM1 no increased prevalence of migraine or any other primary headache was found [7,8] compared to the general population.

There are, however, reports describing a possible relationship between the cerebellum and the pathogenesis of migraine. The cerebellum may be involved in four types of migraine: migraine with aura, migraine without aura, migraine with brainstem aura, and familial hemiplegic migraine. Purkinje cells express calcium channels and may therefore be related to the pathophysiology of hereditary forms of migraine. Calcium channel dysfunction may lead to cerebral and cerebellar hyperexcitability that may facilitate the cortical spreading depression wave [2].

Although CM1 is considered to be a congenital malformation, in our patients symptom onset was in adulthood (17 to 70 years) in agreement with the literature, as was the net female sex predominance (80%) [1].

The main aim of surgery in those patients in whom the headache is found to be associated with CM1 is to reestablish the CSF flow in the subarachnoid space at the level of the craniospinal junction [16]. The patient is placed in the ventral decubitus position with the neck flexed. The head of the table is elevated 30 degrees to decrease the venous pressure. A suboccipital incision is made in the mid line, the posterior part of the atlas is resected, followed by duroplasty in order to provide sufficient subarachnoid space at the level of the foramen magnum, so that the symptoms and signs of the CM1 are reversed [17]. Possible complications of the surgery described in the literature to occur in around 20% of the patients are: pseudomeningocele (30%), aseptic meningitis (9%), cerebellar ptosis, craniocervical

Table 3: Characteristics of the headache in patients with Chiari type I malformation and headache.

QUALITY	
Oppressive pain	56%
Pulsatile pain	39%
Worsening with Valsalva	15 %
DURATION	
Prolonged (more than 3 hours)	85%
Short lasting (less than 3 hours)	15%
INTENSITY	
Severe	84%
Moderate	10%
Mild	6%
ASSOCIATED SYMPTOMS	
Photophobia and/or Phonophobia	85%
Nausea	75%

instability, and recurrent syringomyelia (10 to 40%) [18,19]. No postsurgical complications were observed in our patients who underwent surgery. Outcome was good and headache improved.

CONCLUSION

In patients with recurrent headaches and evidence of CM1, the diagnosis of the type of headache is a challenge.

In the majority of our patients, the finding of CM1 on brain MRI was incidental and headaches were not related to CM1 but were most often headaches due to analgesic abuse followed by different types of primary headaches. Based on this observation we suggest that all patients with CM1 should be evaluated by a neurologist to rule out other causes of the headache and avoid unnecessary surgery.

Interdisciplinary evaluation of these patients in a team consisting of neurologists and neurosurgeons is important to determine the most appropriate therapeutic approach.

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