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CLINICAL ASPECTS OF HEADACHE

Chiari malformation-related headache: outcome after surgical treatment

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Abstract The outcome of headache in a series of 135 operated CM1 is presented. Favorable results were obtained in 85% of atypical and 93% typical headache with the support of a multidisciplinary approach that restricted the indications for surgery.

Keywords Cerebrospinal fluid · Chiari 1 malformation · Headache · Occipital-cervical decompression · Syringomyelia

Introduction

Chiari 1 malformation (CM1) is defined as cerebellar tonsils descend of at least 5 mm into the spinal canal below the plane of the foramen magnum. The brainstem may be occasionally involved; if bony anomalies of the craniovertebral junction are associated as well, the picture is defined CM 1.5. In many instances, tonsil herniation is an incidental finding on neuroradiological imaging and, if asymptomatic, surgical treatment is not necessary. The prevalence in general population is 0.6–0.9% [1]. On the contrary, CM1 may be associated with syringomyelia, obstructive hydrocephalus, or neurological signs and symptoms [2]; in these cases, surgery is the treatment of choice. In rare cases, CM1 patients may experience syncope [3]. Symptoms usually start in the second-third decade of life, but there is increased recognition of symptomatic cases in children [4–6]. Headache is the most common initial symptom, leading to diagnosis in 25–50% [2]. The malformation may reduce the free pulsatile movement of cerebrospinal fluid (CSF) through the foramen magnum during the cardiac cycle, creating a partially entrapped spinal subarachnoid space and causing high cervical subarachnoid pressure waves, enhancing transmural CSF movement into the spinal cord, which is the pathophysiological base of syrinx. The obstruction of the CSF circulation may likewise be important in the origin of headache. The typical CM1-related headache is usually severe and paroxysmal, induced or exacerbated by Valsalva maneuvers such as laughing, sneezing, and coughing [7], in relation to waves of increased intracranial pressure that impact on cerebellar tonsils. However, many other types of headache have been reported including migraine, tension-type headache, and cluster headache [8, 9] but the role of CM1 in the genesis of these "atypical" headaches is still debated. We present a series of patient that underwent foramen magnum decompression (FMD) for CM1 associated with syringomyelia or symptoms, focusing on the impact of surgical treatment on preoperative headache both typical and non-typical.

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Material and methods

Between 1998 and 2017, 143 adult patients affected by CM1 were admitted at our institution for surgical treatment. The malformation was documented in all patients by MRI imaging scans on the sagittal plane; CM1 was confirmed with a tonsil descent of 5 mm in relation to the basion-ophistion plane. Spine MRI scans were also performed to detect syrinx, and flow study in posterior fossa was done to confirm the

constraint to CSF passage posteriorly. All patients had neurological symptoms attributable to the CM1 (i.e., Chiari-type headache, bulbar, or cerebellar dysfunction) or harbored progressive syringomyelia. Patients with an associated congenital syndrome for skeletal dysplasia, such as Klippel–Feil syndrome, were excluded from this study ($n = 4$). Age at surgery ranged between 19 and 73 years old (mean 40.6 years old) with a female prevalence ($n = 90$). The most common indication for surgery was syringomyelia ($n = 67$), syringomyelia associated to typical-CM1 headache ($n = 30$), typical headache ($n = 27$), bulbar, and cerebellar symptoms ($n = 9$), Four patients presented also hydrocephalus; four other patients were lost at follow-up. Typical headache was defined as occipital or suboccipital pain of short duration (less than 5 min) and provoked by cough or other Valsalva-like maneuvers [7]. All types of headache that did not fulfill criteria for typical-CM1 headache were defined as atypical. Taking in account the global series (135 pts), 97 (71.8%) had a syrinx, 29 (21.5%) had just typical headache, and 9 (6.7%) had neurological signs and symptoms. Regarding to the 97 patients in which surgical indication was due to progressive syringomyelia, 34 of them were also affected by non-typical headache, 64 by typical headache, and the remaining 33 never complained for headache. So the total number of patients complaining for preoperative headache was 93 (70%), the great majority affected by typical headache ($n = 89$) and a small number of atypical ($n = 30$). Short-term (<3 months) and long-term (12 months) outcomes were evaluated. Headache improvement was defined as a reduction in the frequency or intensity of the headache at follow-up. Because the end point of the present study was to evaluate the impact of surgical treatment on preoperative headache, the patients with motor impairments and dysphagia ($n = 2$), ataxia ($n = 2$), ataxia plus dysphagia ($n = 1$), apnea and dysphagia ($n = 1$), limb pain ($n = 1$), visual disturbances ($n = 1$), and tinnitus ($n = 1$) were excluded from the final analysis, due to absence of headache. The final analysis about the results of surgery on headache was therefore performed on 93 CM1-operated patients. The clinical data and demographic details of the series are summarized in Table 1. The surgical treatment by FMD was carried out in the prone position. After occipital bone craniectomy with high-speed drill, the posterior arch of the atlas was routinely removed, but C2 and related muscle attachments were preserved. In all cases, the dura was opened under microscopic vision with a midline linear incision. The arachnoid plane was preserved whenever possible, and tonsil resection or coagulation was performed only when the tonsil extended behind the posterior arch of the axis or in case of arachnoiditis. A dural patch was therefore tightly sutured, widening much more just at the craniovertebral passage. The graft was cut to the size of the dural defect and secured with a monofilament non-absorbable suture. Fibrin glue and different sealant products were applied on the suture to prevent cerebrospinal fluid leak. In four patients presenting an hydrocephalus due tonsil herniation, the ventricular dilatation was treated before the CM1 to prevent CSF leakage due to increased intracranial pressure (ICP); in three of them, the first surgical choice was to perform an endoscopic ventriculocisternostomy and in the remaining one, a ventriculoperitoneal shunt was placed.

Table 1 Patient series ($n = 135$)—surgery and symptoms

Males	45		
Females	90		
Age range	19–73 years		
Mean	40.6 years		
Median	41 years		
	Syrinx	No syrinx	Other symptoms
	97 (72%)	29 (21%)	9 (7%)
No headache 42	33 (24%)	–	9 (7%)
Headache 93			
Typical	30 (22%)	29 (21%)	–
Atypical	34 (25%)	–	–

Results

There was no major morbidity neither mortality.

One hundred thirty-two patients were submitted to FMD and 9 (6.8%) of them had different degrees of CSF collections; 8 were treated conservatively with evacuations and compression bandages. Other postoperative complications included hydrocephalus ($n = 2$) and wound infections ($n = 1$). One case of hydrocephalus had ventriculo-peritoneal shunt (VPS) and one patient with a CSF leak underwent reoperation for re-do duroplasy (0.75%). The global favorable response of preoperative headache to surgery for CM1 in our series was 70%. More in detail, 64 patients who were operated for syrinx also had headache. Of them, 29 (85%) patients out of 34 affected by atypical headache experienced an improvement, 2 had no change, 1 temporarily worsened (due to CSF leak that required a second surgery), while 27 (96%) patients out of 30 with typical headache had an improvement. About the 29 patients operated on just for typical headache, without any syrinx, 27 (93%) of them had an improvement, only one had remained stable, and another one temporary worsened due to the occurrence of postoperative hydrocephalus that required a shunt placement. All the 132 patients submitted to FMD were treated in the postoperative period by a protocol aimed to prevent "relative hypotension" occurrence, by overhydration and prolonged supine positioning with recovery of the upright in a couple of weeks. Despite this precaution, 18 patients experienced prolonged intracranial hypotension syndrome

with orthostatic headache and walking difficulties that solved prolonging overhydration and bed rest for 4 weeks.

Discussion

The clinical presentation of CM1, which typically begins in young adults, can include headaches, visual disturbances, neuro-otological complaints, lower cranial nerve dysfunction, and sleep apnea [5, 6, 10]. Those clinical manifestations are related to direct compression syndromes (brainstem or spinal cord) or to cerebrospinal fluid (CSF) disturbances (hydrocephalus, pseudotumor-like episodes, headache). Headache is the most common symptom, occurring in 30–80% of patients [11]. This is true also in the pediatric population, with a frequency of headache ranging from 15 to 75% [12]. By the way, the patients in which cerebellar tonsil displacement is present report a wild spectrum of headache. The typical headache in CM1 is described as occipital or suboccipital pain, brief in duration and triggered by cough, Valsalva maneuver, or physical activity [7]. Also, different types of headache have been associated with CM1, including intracranial hypotension-like headache, long-lasting headache with cervicogenic features, and continuous headache. Pascual [13, 14] reported that all symptomatic cases of cough headache were associated to CM1, and most of them had also other posterior fossa signs and symptoms. This type of headache is supposed to be a consequence to the compression of C1 and C2 roots by further tonsillar descent occurring during Valsalva-like maneuvers. Moreover, the pain could be the result of dissociation of the craniospinal pressure or due to sudden increase in intracranial pressure caused by obstruction of the CSF [15]. In that kind of patients, headache usually remits or improves after successful treatment of CM1 [7]. It is also important to remember that patients with CM1 have the same risk for the common headache disorders as normal population, including migraine and tension-type headache; if these headache disorders are present, they should not be confused with CM1 headache or be considered as an indication for surgery [16]. Other patients with CM1 may present with signs and symptoms of interruption of CSF flow, brainstem, or cerebellar dysfunction including visual disturbances, dysphonia, dysphagia, sleep apnea, incoordination, and sensory disturbances. Motor and sensory difficulties, pain, and scoliosis may be associated with syringomyelia. According to the literature, there is poor correlation between entity of symptoms and radiological severity of tonsil descent. So it is difficult to attribute radiological findings to different types of headache [17]. Despite many debates about the appropriate technique for surgery, there is a consensus between neurosurgeons about the fact that treatment is indicated just in patients with symptomatic CM1 and progressive syringomyelia. A watch and see management is also a reasonable option in patients with mild, non-disabling symptoms, in absence of neurological deficits. The problem rises with the symptom "headache" is taken into account, for the difficulty to discriminating whether or not it depends on the tonsil descent.

Conservative management is generally proposed for atypical headache rather than for the typical one, with specific occipital–suboccipital pain with significant disability and concurrent cough headache. The decision for non-surgical treatment should be taken in consultation with neurosurgeon, neuroradiologist, and neurologist [18]. Surgery is recommended to patients with signs and symptoms of brainstem or cerebellar dysfunction, large or progressive syringomyelia, or with a poorly controlled, typical CM1 headache disorder [19]. Patients should be considered as surgical candidates if the headache is suboccipital, precipitates by Valsalva-like maneuvers, and is refractory to medical therapy and affect everyday life. Moreover, in patients with non-typical CM1 headache, additional signs or symptoms might indicate surgery, in particular bulbar dysfunction (ocular movement disorder, central sleep apnea, swallowing disturbances) or syrynx. The present series is a selection of highly symptomatic cases, with a restrict indication for surgery, offered just in case of progressive syrinx, disabling typical headache of neurological deterioration. The results of surgery on headache in these selected cases was quite good: 93% of patients with typical headache improved, regardless to the association with syringomyelia or less. By the way, also atypical headache had benefits of surgical treatment, with improvement in 85% of cases, even if the surgical indication was due to progressive syringomyelia. Consequently, some role of symptomatic CM1 in facilitation if not in the genesis of other types of headache may be hypnotized. This study present several important limitations. First, headache is a subjective symptom that is difficult to assess. Second, there were few details on headache because the patients were evaluated just by the neurosurgeon in some steps of their clinical pathway; moreover, the diagnostic criteria for headache related to CM1 require validation [7]. It has been hypnotized that when surgical treatment for CM1 fails, we need to evaluate if tonsil herniation is secondary to other predisposing factors such as raised intracranial pressure associated with idiopathic hypertension or occult craniosynostosis [20]. No cases in our series presented such a picture; on the contrary, many of our patients experienced an intracranial hypotension-like headache, quite different from the headache they had before surgery. The self-limiting duration and the good response to hypotension protocols confirm its genesis: a transient passage of relative intracranial hypotension due to CSF loss along surgery and underproduction in the change from a chronic hypertension due to CM1 to a normal range of intracranial pressure. Based on this, it may be hypnotized that the FMD technique applied in the present series was effective in treating headache related to chronic intracranial hypertension due to CM1. The multidisciplinary approach that

we propose, with the aid of the neurologist, increased the percentage of successful surgery by the mean of a more tight indication, excluding from surgery the patients with incidental CM1 affected by different neurological syndromes and headache responsive to medical treatment.

Conclusions

CM1 patients may present a range of neurological signs and symptoms, including different types of headache. Our study reports that headaches will improve after surgery if the indication is correct. A multidisciplinary approach is mandatory to obtain this result.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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