Chiari I - Idiopathic Intracranial Hypertension Association After Failed Posterior Fossa Surgery. Case Report

Asociación de Arnold Chiari I e Hipertensión Intracraneal Idiopática Tras Cirugía Fallida de Fosa Posterior. Caso Clínico

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ABSTRACT

Objectives: Several papers have been published relating the Idiopathic Intracranial Hypertension Syndrome (HTII) to the Arnold Chiari type I malformation (AC1M). Both entities have clinical and demographic similarities, a poorly defined etiology and, sometimes common therapeutic possibilities. A correlation between both entities has been suggested, especially in a subgroup of patients in whom posterior fossa decompression surgery fails. With regard to a case, we reviewed the literature and proposed our hypothesis about the origin of Chiari-HTII syndrome and its therapeutic possibilities. Case presentation: A 41 year-old patient with mild obesity, menstrual abnormalities and empty Sella Turcicae, was operated on with an AC1M associating basilar impression and syringomyelia causing all together a centromedullary syndrome. After posterior fossa decompression surgery and successful arthrodesis, she improved in the immediate postoperative period. Nevertheless, she soon developed symptoms of intracranial hypertension (ICH), and showed increased opening pressure in lumbar puncture compatible with HTII syndrome. A ventriculoperitoneal shunt (VPS) was implanted with clinical improvement and 12 months later the syringomyelia was absent on in the magnetic resonance (MRI). Conclusion: The Chiari I-HTII syndrome is described as the coexistence of ICH symptoms after failed posterior fossa surgery, in patients with no flow MRI anomalies, and increased opening pressure at the lumbar puncture. In our experience, both entities seem to overlap in a common syndrome and must be taken into account, especially in patients with atypical onset of symptoms or patients in whom conservative treatment fails.

Keywords: Arnold Chiari I; Idiopathic Intracranial Hypertension; Empty Sella Turcica; Basilar Impression; Pseudotumor Cerebri Syndrome; Posterior fossa decompression. Ventriculoperitoneal Shunt

RESUMEN

Objetivos: Se han descrito artículos cada vez con más frecuencia, que relacionan la hipertensión intracraneal idiopática (HTII) con la malformación de Arnold Chiari tipo I (AC1M). Ambas presentan similitudes a nivel clínico y demográfico, una etiología poco definida y un tratamiento en ocasiones común. Se ha sugerido una correlación entre dichas entidades sobre todo en un subgrupo de pacientes en los que la cirugía de descompresión de fosa posterior fracasa. A propósito de un caso, revisamos la literatura a este respecto y planteamos nuestra hipótesis acerca del origen del síndrome Chiari-HTII así como las posibilidades terapéuticas a este respecto. CASO clínico: Presentamos una paciente de 41 años, con antecedentes de obesidad leve, alteraciones menstruales y una silla turca vacía, ...
The Arnold Chiari type I malformation (AC1M) is defined anatomic and radiologically as a descent of the cerebellar tonsils from 3 to 5 mm through the foramen magnum towards the cervical spine. It has a poorly defined aetiology, however a pathology secondary to a skull-brain disproportion is assumed. Theoretically, there is an increased pressure secondary to any increase in the brain volume or decrease in the size of the skull (such as a small posterior fossa) that causes a secondary descent of the cerebellar tonsils, causing all together the aforementioned pathology. There are also numerous studies that attribute the aetiology to an alteration in the cerebrospinal fluid (CSF) flow that causes a differential pressure between the brain and the spinal cord, which would explain the syringomyelia frequently associated. Clinical symptoms usually associate headache and cervicalgia. The surgical treatment commonly performed is posterior fossa decompression (PFD) in order to increase the relation between the continent- (skull) -content (brain, blood, CSF).

The Idiopathic Intracranial Hypertension (HTII) syndrome is defined as the increase in Intracranial Pressure (ICP), with an increase in the CSF opening pressure in lumbar puncture greater than 25 cmH₂O (19 mmHG) in the absence of ventriculomegaly occupant lesions or infection. Radiologically, the syndrome can coexist and be accentuated in the case of an empty sella turcicae (also related to alterations in CSF and ICP dynamics) or alterations in the craniocervical junction (platibasia or basilar impression), since they add an additional component of instability. The pathophysiology of HTII is also unclear, but it is believed to be secondary to CSF circulation disorders. The most common clinical symptoms are headache, retrobulbar pain and visual disturbances. The typical sign is papilledema. Treatment options vary from diuretics such as furosemide or acetazolamide to more invasive measures such as the implantation of shunt systems, endovascular techniques or even decompression surgery.

There are many similarities among AC1M and HTII. Both present a similar clinical spectrum (headache, sensory motor alterations, diplopia, axial and retrobulbar pain). They also share a pathophysiology with alterations in CSF flow dynamics, demographical similarities, both being very prevalent in obese middle-aged women. Lastly, they both partially respond to decompression surgeries.

Associations between these two entities have been described in numerous scientific articles. However it is not clear if it is due to a coincidence (since the advent of the MRI more cases of cerebellar tonsil descent are diagnosed), or if it is due to a syndrome that includes CSF flow anomalies (which would also include syndromes such as the empty sella turcicae, as in our case) or even a cause-effect relationship (postulated by Bejiani) in which an increase in ICP causes a secondary...
descent of the cerebellar tonsils. It has also been suggested a blockage in the circulation of CSF at the foramen magnum that secondary causes an increase of intracranial pressure (ICP)\textsuperscript{3,6,19}.

The association of AC\textsubscript{1}M and HTII has been described in up to 2.7\% of the cases according to different series\textsuperscript{5,11,14,15,17,18}, as well as after failed posterior fossa decompression\textsuperscript{1,14,15,16,17,18}. We pose the possibility of a coexistence of both entities in the same clinical syndrome. We carried out a complete review of the literature with the aim of optimizing the treatment possibilities of these patients and with the future expectation to perform studies of greater scientific evidence that serve as a guide in the therapeutic management of this pathology.

We present a 41-year-old woman with history of mild obesity (Body Mass Index [BMI] 26.3), menstrual irregularities, and non-smoker. She was referred to the external consultations of Neurosurgery at our institution presenting clinical symptoms compatible with centromedular syndrome, of around 13 months.

The MRI showed tonsil descent greater than 5 mm, compatible with an AC\textsubscript{1}M, associating basilar impression, syringomyelia and perilesional oedema. At the supratentorial compartment, an empty sella turcicae with ventricles of normal size and morphology was visualized.

The patient was surgically approached through a posterior fossa decompression, with additional resection of the posterior arch of the atlas and secondary occipito-C3 arthrodesis to treat the instability associated with basilar impression. It was performed a layer-by-layer closure with artificial duraplasty. The patient immediately improved the previous symptoms and was discharged from the hospital.

After 20 days she developed headache and incipient nausea with morning predominance. At physical examination we observed a tension wound compatible with a contained CSF fistula, with no CSF leak, nor fever. Urgent analysis did not show signs of infection.

Computed tomography (CT) and craniocervical MRI were performed to confirm radiologically the initial clinical suspicion and a surgical reintervention of revision and duraplasty, associated with the implantation of an external lumbar drainage was performed.

Progressively the patient improved from headache, with the external lumbar drainage (ELD) draining about 500cc of CSF per day. Both the CSF analysis and the cultures were repeatedly negative. However, it drew attention in the hormonal series, the prolactin increase of about 400 as well as increase in headache when attempting to decrease the drainage volume of CSF. In subsequent brain CT scans there was no increase in ventricular size despite the symptoms of intracranial hypertension (ICH). There were no associated visual deficits or clinical signs of infection.

After a thorough study of the scientific literature and according to the clinical and radiological context of the case, a ventriculoperitoneal shunt (VPS), (Hakim Adams, Codman; Los Angeles, California, USA), neuronavigation-guided (Stealth Station, Medtronic; Fridley, Minnesota, USA), was implanted.

The patient improved clinically, showed no signs of CSF fistula and was discharged to the external consultations of our department, where she is followed ever since. In the control MRIs, CSF accumulation was not observed and syringomyelia disappeared progressively.
DISCUSSION

The AC1M’s clinical spectrum is very broad. There have been described symptoms related to brainstem compression and lower cranial nerve paresthesias, cerebellar disturbances, cervical and centromedullary alterations\(^1\). Also increased ICP, headache and cervicalgia have been described. However, there have been recently attributed to the AC1M atypical symptoms such as migraine, retrobulbar pain or dizziness, as well as a relationship with other syndromes such as multiple sclerosis, fibromyalgia, chronic fatigue syndrome, and even otological diseases such as Menière’s syndrome\(^2\).

In 30% of the cases, AC1M represents an accidental finding in imaging tests, especially since the onset of MRI (0.77% of the cases the MRI presents a descent in cerebellar tonsils)\(^2\). The syndrome is more prevalent in women (66%) and presents an incidence of 0.5% in the European population. The classic treatment is posterior fossa decompression surgery.

The most frequent clinical manifestations in the HTII are headache, transient visual disturbances, tinnitus, photopsia and retrobulbar pain. The typical sign is papilledema, which, sustained over time, can cause secondary atrophy of the optic nerve. However, motor sensory disturbances, dizziness, acral paresthesia, chronic fatigue syndrome and axial pain have been included within the spectrum of HTII, similar to those described in the AC1M\(^5\).

The CSF flow dynamics is due to the interaction of the vascular system, cerebrospinal fluid and cerebral parenchyma within a closed and non-distensible cavity such as the skull. Some authors describe an increase in ICP that influences the capacity of CSF absorption by the cerebral venous system\(^\ref{5,19}\). Sahs and Joint\(^15\), in 1956, revealed through anatomopathological studies an increase in interstitial oedema in patients with HTII. Johnston et al.\(^12\) described a decrease in the gradient pressure between the subarachnoid space and the superior sagittal sinus by isotopic cisternography. According to the working hypothesis, pressure in the venous system increases as the CSF accumulates in the subarachnoid space, until a pressure gradient capable of overcoming the vascular resistance is created, thus producing the passage of CSF to arachnoid granulations or until the pressure in the subarachnoid space exceeds that of the superior sagittal sinus.

Moser\(^*\) demonstrated by means of MRI diffusion sequences an increase in white matter oedema, which would explain the small ventricular size.

Bejani\(^\ast\), on the other hand, attributed the HTII to an increase in brain volume in relation to the other two compartments and postulated a cause-effect hypothesis with the AC1M, in which the amygdala descent is due to an increased intracranial pressure secondary to an increased parenchyma volume.

Up to 90% of patients with HTII are obese women aged between 20 and 44 years old\(^1,3,8,10,12\). Treatment varies from diuretics such as furosemide and acetazolamide to more invasive measures such as the lumbo-peritoneal shunt system (LPS), considered gold standard, ventriculo-peritoneal shunt system (VPS), endovascular techniques, and even suboccipital and subtemporal decompressions. Cranietomies produce rapid relief through alteration in the compliance between the skull, CSF and brain tissue\(^1,3,11,15\). However, many authors believe that scarring, especially of the dura mater, decreases elasticity in the medium term due to fibrous scar tissue, causing surgery failure\(^1\). Likewise, surgery is effective mainly for treating papilledema and preventing secondary optic atrophy, but it does not improve headache or neck pain\(^1,5,11\).

Chiari I-HTII syndrome\(^2,11,15,16,17,18\) is described as the coexistence of common signs and symptoms in a suitable radiological context. The aetiology of both can correspond to the coexistence in the same syndrome or to be a consequence one of the other. Also, patients with failed posterior fossa surgery of AC1M have been diagnosed posteriorly of HTII\(^5,15,16,17\).
We do not believe that such entities have a coincidental association. Johnston et al. described an association in MRI up to 6% of both entities, being eight times more frequent than the tonsil descent greater than 5mm described in 0.77% of MRI performed by other causes. Nor do we believe that it is due to a CSF blockage at the foramen magnum, since the clinical response to the external lumbar shunt goes against an obstruction, in which symptoms would worsen.

Therefore, we consider that both pathologies can overlap in a common syndrome. In some patients with clinical symptoms and MRI compatible with AC1M, a posterior fossa decompression surgery is performed. They improve temporarily by increasing brain compliance, but after the fibrous tissue of the scar of the duramater, the cerebral distensibility decreases rapidly, thus reversing the pulse pressure curves and giving symptom exacerbation, in a period of time that oscillates between six and twelve months. In the series reported by Bejjani and by Fagan, up to 46% of patients with failed posterior fossa decompression surgery in the context of a AC1M presented an association of HTII. It is noteworthy that in the Bejjani series patients obtained satisfactory responses to shunt systems, while in the Fagan series it failed in most adult patients (in children it was effective). It is also true that both series presented a number of patients too small to draw significant statistical conclusions.

The possibility of performing a lumbar puncture has been suggested in patients diagnosed with tonsil descent so as to try to simultaneously diagnose both pathologies and to be able to optimize the treatment possibilities. Likewise with the ICP pressure monitoring systems both entities can be diagnosed within the same admission lapse of time, by performing MRI and measuring the ICP with an intraparenchymal device.

In our particular case it may correspond to a true association of both syndromes, due to AC1M, which dramatically improved with surgery, but coexisting with an HTII that showed up posteriorly at an early stage and also improved after the implantation of a VPS.

Furthermore, it could correspond to a common syndrome that showed up after posterior fossa decompression surgery and could have been managed with a single surgery such as the VPS. Hypothetically the VPS would first resolve the ICH and secondarily the ACM1. But again, this hypothesis needs further investigation with studies of higher level of evidence.

CONCLUSION

The Chiari I-HTII syndrome is described as the coexistence of ICH symptoms after failed posterior fossa surgery, in patients with no flow MRI anomalies, and increased opening pressure at the lumbar puncture. In our experience both entities seem to overlap in a common syndrome and do not correspond to a cause-effect relationship, since the patient presented early clinical worsening after the first surgery and immediately improved with high volumes of CSF drainage and thus, the implantation of a VPS.

The monitoring of ICP in patients in whom this association is suspected, as well as the realization of studies with greater level of scientific evidence could help us in the future clarify the doubts to this respect and to be able to accomplish a better therapeutic approach to these patients. Ideally this syndrome could even be solved with an only surgical intervention, but this, again, must be further investigated.
It is noteworthy that in the Bejjani series patients obtained of patients with failed posterior fossa decompression surgery. In the series reported by Bejjani and by Fagan1,16, up to 46% of patients with tonsil descent greater than 5mm described in 0.77% of MRI performed by other causes. Nor we do believe that it is due to a CSF blockage at the foramen magnum, since the pressure curves and giving symptom exacerbation, in a period of time that oscillates between six and twelve months. It is suspected, as well as the realization of studies with greater level of evidence.

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Figure 1. Pre and postoperative images, obtained from T2-weighted MRI. A. Preoperative T2-MRI showing an Arnold Chiari type I lesion, with cervico-thoracic syringomielia and basilar impression. B. Post operative T2-MRI showing the improvement of the amigdalar herniation and the syringomielia after surgery.

Figure 2. A-C Preoperative Images obtained from T2-weighted MRI (A,B) and 3D CT reconstruction (C). A. T2-MrRI with normal sized ventricles, compatible with Intracranial Idiopathic Hypertension Syndrome. B. T2-MRI associating an empty sella turcicae. C. 3D CT reconstruction showing the association of basilar impression.
Figure 3. Postsurgical images after first (A, B, C) and second (D) surgery. A-B. Coronal and Sagittal 3D CT reconstruction after first surgery; C. Sagittal postoperative CT after first surgery showing occipito-C3 arthrodesis; D. Postoperative CT after second surgery. Navigated Ventriculoperitoneal shunt with the tip of the catheter located in the foramen of Monroe.

Figure 4. A-D Intraoperative microsurgical images. A. Left cerebellar amygdala subarachnoid dissection; B. Left cerebellar amygdala after complete liberation; C. Both cerebellar amygdala fully liberated and opening of the fourth ventricle; D. Final stage after duraplasty sutured with 7.0 nylon.
REFERENCES


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