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EXPERIENCE REPORT

Chiari Malformation: experience report

Malformación de Chiari: relato de experiencia

Malformação de Chiari: relato de experiência

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Abstract

The aim was to produce an experience report about a patient with several disorders related to Chiari syndrome type I, also addressing other findings found in her. The patient described in the case was previously diagnosed with Arnold-Chiari Malformation type I, which is characterized by the existence of an ectopia of the cerebellar tonsils, which are located underneath the foramen magnum. Such a patient meets all clinical, laboratory, and imaging criteria for this pathology. Type I Chiari Malformation is the projection of the cerebellar tonsils towards the foramen magnum. This projection compresses the brain stem and spinal cord; Therefore, the cranial nerves are affected. It is worth mentioning that there is partial or total obstruction of the cerebral aqueduct. It is concluded that Chiari I, as it affects the cerebellum and adjacent structures, brings general symptoms, which makes diagnosis difficult. Efforts must be aimed at improving effective surgical techniques that reduce complications and sequelae for the patient.

Descriptors: Arnold-Chiari Malformation; Hydrocephalus; Abnormalities; Cerebrospinal Fluid; Magnetic Resonance Imaging.

Resumén

El objetivo fue realizar un relato de experiencia sobre una paciente con varios trastornos relacionados con el síndrome de Chiari tipo I, abordando también otros hallazgos encontrados en ella. El paciente descrito en el caso fue diagnosticado previamente con Malformación de Arnold-Chiari tipo I, la cual se caracteriza por la existencia de una ectopia de las amígdalas cerebelosas, que se ubican debajo del agujero magno. Un paciente así cumple con todos los criterios clínicos, de laboratorio y de imagen para esta patología. La malformación de Chiari tipo I es la proyección de las amígdalas cerebelosas hacia el agujero magno. Esta proyección comprime el tronco del encéfalo y la médula espinal; por tanto, los nervios craneales se ven afectados. Cabe mencionar que existe obstrucción parcial o total del acueducto cerebral. Se concluye que Chiari I, al afectar el cerebelo y estructuras adyacentes, trae síntomas generales, lo que dificulta el diagnóstico. Los esfuerzos deben ir dirigidos a mejorar técnicas quirúrgicas efectivas que reduzcan las complicaciones y secuelas para el paciente.

Descriptores: Malformación de Arnold-Chiari; Hidrocefalia; Anomalías; Líquido Cefalorraquídeo; Imagen de Resonancia Magnética.

Resumo

Objetivou-se produzir um relato de experiência sobre uma paciente com vários acometimentos referentes a síndrome de Chiari tipo I, sendo abordado também outros achados encontrados na mesma. A paciente descrita no caso foi previamente diagnosticada com a Malformação de Arnold-Chiari tipo I, a qual se caracteriza pela existência de uma ectopia das amígdalas cerebelares, que se situam por debaixo do forame magno. Tal paciente tem todos os critérios clínicos, laboratoriais e de imagem para esta patologia. A Malformação de Chiari tipo I é a projeção das amígdalas cerebelares, em direção ao forame magno. Essa projeção comprime o tronco encefálico e a medula espinhal; portanto há o acometimento dos nervos cranianos. Vale ressaltar, que há obstrução parcial ou total do aqueduto cerebral. Conclui-se, que o Chiari I, por acometer o cerebelo e estruturas adjacentes, traz sintomas generalistas o que dificulta diagnóstico. Os esforços devem se destinar ao aperfeiçoamento de técnicas cirúrgicas eficazes que diminua as complicações e sequelas ao paciente.

Descritores: Malformação de Arnold-Chiari; Hidrocefalia; Anormalidades; Líquido Cefalorraquidiano; Imageamento por Ressonância Magnética.

Introduction

The first publication on what would later be called Chiari Malformation was published by Hans Chiari (1851–1916) in an Austrian magazine, the Deutsche Medizinische Wochenscriffem in 1891 with the title "Concerning alterations in the cerebellum resulting from cerebral hydrocephalus". The first type described, which came to be known as Chiari Type I, was characterized by elongation of the tonsils and medial divisions of the lower lobes of the cerebellum into cone-shaped projections that accompany the medulla oblongata in the spinal canal. Five years later, Chiari speculated that these changes could be related to the defective formation of the craniovertebral junction, which in Brazil was correlated with a high prevalence of the syndrome in the Northeast of the country due to the predominant brachycephalic profile in this region. All cases, until then, were related to congenital hydrocephalus or not. In 1981, the first case of type I Chiari Malformation unrelated to hydrocephalus was described. Another researcher, Arnold, even described this malformation associated with spina bifida, which resulted in the term "Arnold-Chiari Malformation". However, the contribution was negligible, which culminated in the designation of just "Chiari Malformation" and its types. It was during the 1970s that the term adult Chiari malformation gained popularity and large studies were disseminated[1-3].

Arnold-Chiari malformation is a congenital alteration of the hindbrain, where part of the cerebellar cortex is depressed posteriorly through the spinal canal, together with herniation of the spinal cord and fourth ventricle into the cervical canal. Four types have been described, presented below, with the most prevalent form being Chiari type I, as per the description: Type I, caudal prominence of the cerebellar tonsils, reaching at least three millimeters below the foramen magnum, is rarely observed below the second cervical vertebra, which may eventually be associated with hydrocephalus. Type II, in the herniation of the tonsils, cerebellar vermis, lower portion of the medulla oblongata, and IV° ventricle, through the occipital foramen. Type III, in herniation of the cerebellar vermis and contained brainstem of a high cervical meningocele. Type IV, severe cerebellar hypoplasia or aplasia, is associated with the posterior fossa with little evolution[4-6].

Chiari malformation is an uncommon pathology, with women being most affected (80% of patients). It is a pathology that accounts for between 1 and 4% of all neurosurgical alterations. Determined according to the protrusion of the cerebellar tonsils, a minimum of 3mm under the foramen magnum in the case of adults and 6mm in the case of children. Recognition is determined months and even years from the onset of symptoms and is proven in all cases (100%) through magnetic resonance imaging (MRI). Protrusion of tonsils within 3mm and 5mm is termed as mild and non-progressive cerebellar ectopia and needs connection along with medical and radiological findings. The intervention is surgical in patients with indicative symptoms, equivalent to a discussion of the real need in those who present few symptoms. Surgical intervention has a low mortality rate ranging from 0 to 0.5%[7,8].

Hydrocephalus usually accompanies this pathology and is divided into communicating, non-communicating, and constrictive. Communicating lesions are caused by defects in drainage to the subarachnoid space. Non-communicating disorders are related to obstruction of CSF drainage sites and, finally, hydrocephalus caused by the constrictive hypothesis, which is related to Chiari pathology, which will cause the cerebellum, with its swelling, to compress the brain stem, preventing CSF drainage into the subarachnoid space, leading to the formation of hydrocephalus. Therefore, hydrocephalus related to Chiari Type I is called "Obstructive Hydrocephalus", which is related to the obstruction of the foramina of Magendie and Luscka, preventing CSF flow to the subarachnoid space and increasing the amount of CSF in the intracranial cavities, thus generating intracranial hypertension and dilation of the cerebral ventricles. The known prevalence of the anomaly is 1 in every 1,000 to 5,000 people[4,9-12].

The objective of this study is to address the case of Chiari malformation in detail, showing the disorders chronologically so that there is a greater understanding and comprehension of this pathology. Furthermore, the aim is also to analyze the treatments carried out on such a patient, so, that there is an understanding of the effectiveness.

Methodology

A descriptive study, of the experience report type, was carried out to understand a patient, through retrospective analysis and follow-up of the diagnosis of Chiari type I malformation related to hydrocephalus on head CT scans. The patient's gender, age at diagnosis, obstetric history, and general pathologies were recorded. Based on the patient's history, the medical history was organized chronologically to better understand the natural history of the disease.

Experience Report

Female patient, 18 years old, with Chiari type I and obstructive hydrocephalus, firstborn. During her pregnancy at 37 weeks, through ultrasound, she noticed the presence of dilated lateral ventricles, with a VL/HC ratio of 57 (normal for 37 weeks: 24-34). She had some segments measured at birth; these are the biparietal diameters: 91mm; femur length:71mm; waist circumference: 335 mm; head circumference: 333mm; approximate fetal weight: 3,180 grams; approximate height: 42cm; biophysical profile: 8/8. By measuring the head circumference, the presence of hydrocephalus was confirmed.

On April 24, 2001, a computed tomography examination of the brain segment was requested, which showed obstructive hydrocephalus, with a pattern of aqueductal stenosis and enlargement of the intermediate mass. The other day, the patient was requested to be hospitalized because she had a slightly bulging anterior fossa (AF), with a PC of 41.5 cm, and was then submitted to neuroendoscopic III ventriculostomy. Two more III ventriculostomies were later performed, in September 2017 and June 2018.

On September 19, 2003, the patient was diagnosed with angiotoma lesions (lesions caused by bacteria, on the surface of the skin, generally in immunocompromised people) on the back and left lower limb (material sent for biopsy; the sections showed nodular melanocytic proliferation); the sections show skin showing a benign dermal lesion, with exuberant vascular proliferation, characterizing a hemangioma.

On October 16, 2007, the patient underwent a magnetic resonance imaging (MRI) scan at the Axial Imaging Center and found: Partial agenesis of the septum pellucidum (atrophy of the septum pellucidum in its embryonic phase), the 3rd ventricle presented with relative deformity, resulting from partial fusion of the thalami. 4th ventricle of normal dimensions and anatomical configuration. Due to agenesis of the septum pellucidum, partial fusion of the thalamus in the midline was observed. He also reported some focal images suggestive of heterotopic gray matter adjacent to the frontal horn and horn of the left lateral ventricle, in a subependymal situation; focal areas suggestive of heterotopic gray matter in the subependymal region adjacent to the occipital horn of the right lateral ventricle. An appearance suggestive of closed-lip schizencephaly was found - closed-lip schizencephaly is a disease that is characterized by having fissures from the pial surface to the ependymal surface, with the edges covered in gray matter, in this case, the schizencephaly is lip closed, that is, the two cortices are juxtaposed and without the presence of CSF - in the region of the right frontal convexity. It was not possible on that date to study the CSF flow, due to the marked aqueductal stenosis. There was dilation of the supratentorial ventricular system, resulting in aqueduct stenosis: there was no evidence of transpedimal CSF transudation.

On October 20, 2010, the patient underwent an MRI of the right forefoot, and a solid expansive lesion was observed in the middle of the muscle bellies of the 3rd plantar layer, in the first digital interspace at the height of the middle thirds of the diaphysis of the first and second metatarsal bones, origin clarify. In the differential diagnosis, possibilities such as hemangioma or possibly neurinoma must be considered. Anatomopathological finding: Right plantar tumor, microscopic findings revealed a fragment of fibroadipose, and skeletal muscle tissue partially occupied by a well-defined benign neoplasm, with morphology corresponding to cavernous hemangioma, also reported absence of mitoses, cellular atypia, or infiltrative signs. Ten days later, surgery was performed to remove the tumor from the right foot.

On November 12, 2013, the patient was in a day and maternity hospital and was diagnosed with neuropathy and bone deformities in the right forefoot, which again had to undergo a surgical procedure to remove the tumor in the right forefoot.

On December 23, 2015, the patient went to the BH Orthopedic Hospital with pain in her left foot, where the neuroma (thickening of the tissue around a nerve) was removed. In the last MRI, she presented a residual lesion from the resected tumor and may be showing new growth of the tumor lesion. The use of medication and closed shoes, such as sneakers, were tried to improve the pain. Walking should be limited, with pain being the limiting factor. On December 28, 2015, she had a recurrence of pain in her right foot, which was also subjected to an MRI.

In a control examination in 2016, he reported the following aspects: septo-optic dysplasia (Morsier's disease), cortical dysplasia in the anterior aspect of the cingulate gyrus on the right and cranial part of the parieto-occipital sulcus on the left, hypoplasia of the frontal lobes as well such as cerebral falx related to enlargement of the inter-thalamic adhesion, admitting overlap with lobar holoprosencephaly and supratectorial ventriculomegaly with a pattern of compensated stenosis of the caudal third of the aqueduct, from an imaging point of view, maintaining the same appearance as in the last examination mentioned above. In August of the same year, the patient was admitted to Belo Horizonte under a neurosurgical specialty complaining of headache and neck pain, when she underwent an MRI of the skull and cervical spine showing hernination of the cerebellar tonsils at the level of the posterior arch of C1. Chiari 1 malformation was first described in the patient. As a treatment, PO decompression of the posterior fossa was performed, progressing well without complications. In November, the patient in the late postoperative period of craniocervical decompression, recurrent visits to the emergency room lucid, euphasic, symmetrical and photoreactive pupils, cerebellar tests without changes, preserved oculomotricity, gait without ataxia with support with complaints of headache, fever, dizziness associated with nausea (no improvement after administration of medication).

The patient underwent a two-dimensional echocardiogram and pulsed, continuous, and color Doppler. Images were obtained through the parasternal, apical, suprasternal, and subcostal windows. Regarding the interatrial septum: it was observed that it was intact via the transthoracic approach. The interventricular septum was intact. The atrioventricular and arterial ventricle connection was concordant. Regarding the atria: the left and right atrium had normal intracavitary dimensions with no sonographically visible thrombi. The right and left ventricles have normal intracavitary dimensions. Its walls have normal thickness with global and segmental contractility preserved. LV systolic function indices are within normal limits. The infundibular region and the LV outflow tract are free, without obstructions. Preserved diastolic function of the ventricles. The mitral and tricuspid valves have a normal morphological appearance, with preserved leaflet openings. Laminar mitral flow. Mild tricuspid regurgitant flow. Pulmonary arterial systolic pressure with an approximate value of 23 mmHg for right atrial pressures of 5 mmHg. The aortic valve has a normal trivalvular appearance, with a preserved leaflet opening. Absence of aortic regurgitation or stenotic flow on continuous, pulsating Doppler. Pulmonary valve with normal appearance and movement, both on two-dimensional examination and on pulsed and color Doppler. Normal systolic flow in the ascending aorta and pulmonary artery. Regarding the large vessels, the aortic root and aortic arch were normal. The pulmonary artery trunk was of

normal caliber and the pulmonary branches were confluent. The pericardium had a normal morphological appearance with no effusion. Therefore, ECG with color Doppler: showed mild tricuspid regurgitation reported as physiological.

In 2017, a genetic report on the patient was carried out in Belo Horizonte. She is the only daughter of a healthy, non-consanguineous couple, now separated. Her mother is 44 years old, and her father is 43 years old. Her father has three healthy children from another relationship. There is no other relevant data in the family history. At 37 weeks into her pregnancy, hydrocephalus was noticed. There are no reports of exposure to teratogens. Full-term birth (37 weeks), cesarean section. Apgar 8 and 9. Weight 2,985g, length 47.5 cm, head circumference 34 cm. At birth, marked ventriculomegaly and aqueduct stenosis were confirmed and a ventriculostomy was performed. Later he presented with dental dysplasia, hearing loss on the right, and growth retardation. Subsequent magnetic resonance imaging revealed, in addition to ventriculomegaly, neuronal migration anomalies, with heterotopic gray matter, subependymal nodules, hypoplasia of the frontal lobes, caudal projection of the cerebellar tonsils (Chiari type 1), hypoplasia of the corpus callosum, cerebellar vermis and the distal third of the infundibulum. Normal female karyotype, 46, XX. She had a firm neck at six months, sat at seven months, walked at two and a half years, -she spoke at one year-. He started school when he was one and four months old and did well until the fifth grade when he started to have difficulties. Currently, in the second year of high school, takes separate tests, but has never failed. In his physical examination, he observed a good general appearance, macrocrania, broad forehead, almond-shaped eyes, palpebral fissures slightly inclined upwards, epicanthal folds, nasolabial philtrum little marked, thin upper lip, high palate, flat, everted feet. I did not hear murmurs or palpate visceromegaly. Hybridized genomic comparison array (aCGH) arr (1-22, X) x2 (normal scan). No deletions or duplications were detected. Although his phenotype overlaps with septo-optic dysplasia, I did not find them sufficient to confirm this diagnosis. When she wants children, further genetic counseling will be necessary.

On February 6, 2018, in Belo Horizonte, a computed tomography scan of the chest was performed. The clinical indication was for having juvenile rheumatoid arthritis presenting nodulations in a tomographic examination carried out in August 2017. Volumetric acquisitions were performed in the axial plane using the multislice technique for the chest without the intravenous administration of contrast medium and with a high-resolution standard to the lung parenchyma. The acquisitions were made, one of them during maximum inspiration, with the patient in the prone position, and the other during forced expiration, with the patient in the supine position. In post-processing, multiplanar reformatting was carried out. The observations made by the doctor were as follows: "ground glass" nodular opacities visible in the lower lobe of the right lung in an examination dated August 2, 2017, are not present in the present study.

On June 19, 2018, the patient underwent another hospitalization. Again, the reason was hydrocephalus, which was subsequently treated with an endoscopic third ventriculostomy, progressing to headache. Radiological follow-up with skull MRI revealed ventricular dilation and third ventriculostomy stenosis. She was admitted for revision of the III VT on an elective basis. Good postoperative evolution, without complications. He was discharged in good general condition on June 21, 2018. He received recommendations such as scheduling a return visit, correctly using prescribed medications, and removing the stitches within 15 days.

On July 2, 2018, an MRI study was carried out on the brain. Some aspects were observed, such as there are signs of a ventricular puncture path in the right frontal lobe (access for III ventriculostomy), with an area of hypersignal on T2 and FLAIR, with contrast uptake. Identifies the presence of a flow signal communicating the third ventricle with the suprasellar cisterns. The lateral ventricles are enlarged and asymmetrical, with the left lateral ventricle being larger than the right. The III and IV ventricles have reduced dimensions. There are signs of stenosis of the cerebral aqueduct. there is no evidence of transependymal CSF permeation. The corpus callosum is stretched due to ventricular dilation. The brain stem is preserved. The medical impression was as follows: Postoperative changes (recent III ventriculostomy, suboccipital craniectomy, and decompressive C1 laminectomy); Asymmetric dilation of the lateral ventricles; Cerebral aqueduct stenosis; Presence of flow signs communicating the third ventricle with the suprasellar cisterns and median retro-cerebellar cystic collection, at the site of the suboccipital craniectomy, unchanged concerning previous exams.

On July 5, 2018, the patient underwent an ophthalmic examination with signs and symptoms (amaurosis fugax, numbness, nausea) but there were no extrinsic changes. Glasses were prescribed and retinal mapping was requested.

On July 14, 2018, the patient underwent a multislice CT scan of the skull/brain. The study was carried out and released urgently, using a multislice device, through the volumetric acquisition of images in the axial plane, guided by digital radiography, without the intravenous administration of iodinated contrast medium. Some aspects were observed, such as a comparative assessment carried out with a previous examination dated April 9, 2018. Once again, the stigmata of occipital decompressive craniectomy and previous ventricular shunting in the right frontal region can be seen. An extra-axial collection also persists in the topography of the occipital surgical site, with a density like that of cerebrospinal fluid, measuring 6.7x2.0x4.6 cm (W x AP x T), with characteristics and dimensions similar to those observed in the previous examination on 9 April 2018 (average 7.0 x 1.9 x 4.6cm). The marked asymmetric increase in the dimensions of the supratentorial ventricular compartments remains without evidence of ependymal transudation, similar in appearance to that observed in the previous study of April 9, 2018. The fourth ventricle had the usual morphology and dimensions. The basal cisterns of

reduced dimensions. There was a reduction in the amplitude of the intergyral grooves. The brain parenchyma with usual attenuation coefficients for white and gray matter. The brain throne and cerebellar parenchyma without abnormalities are detectable by the method. Low cerebellar tonsils. Agenesis of the left frontal sinus (anatomical variation) and paranasal sinuses visualized with preserved radiological transparency (non-directed examination). Therefore, the medical impression reported was the absence of significant changes evident in the previous study on April 9, 2018.

On August 10, 2018, the patient underwent hospitalization, with persistent headaches over the last few days. The patient underwent valve implant surgery (PVD). After the NDN exam was performed, the surgery took place on August 10, 2018, without any complications during the procedure. At discharge, the patient was diagnosed with Hydrocephalus/ PO PVD. Hospital discharge was recommended, with a date set for the removal of the stitches on August 20, 2018. The patient had supervised outpatient follow-up, to analyze and guide alarm signs and return if necessary.

In September 2019, a total abdominal ultrasound was performed, which showed that the liver, bile ducts, pancreas, spleen, and right and left kidneys had normal dimensions without any anatomical or functional changes, as well as the abdominal aorta, bladder, and abdominal and pelvic cavity. Another aspect observed was the gallbladder, which has normal dimensions and regular walls, presenting a small echogenic oval formation, fixed in its anterior region, measuring 4.1 mm. Bile has a clear appearance. The impression showed mild hepatic steatosis, and an image suggestive of a polyp in the gallbladder.

In the ECG performed during wakefulness and spontaneous drowsiness, with a test of hyperventilation, carried out in March 2019, a discrete, moderately organized base activity was found, associated with moderate dysfunctional sleep signs, of a non-specific, diffuse nature.
In March 2019, in consultation with a rheumatologist, juvenile rheumatoid arthritis was revealed, which was asymptomatic from a joint point of view, and was advised to undergo follow-up. Recently, in April of this year, a hearing assessment revealed profound sensorineural hearing loss on the right CID H90-5, and the use of a prosthesis for sound amplification was indicated.

Discussion

The predominance of clinical histories of Arnold-Chiari type I malformation is much more evident among young people, with an incidence and predominance of 92% of reports, as symptoms tend to be asymptomatic over several years[5].

Headache is the symptomatology that is equally general in the malformation and can be explained by occlusion of the movement of the spinal CSF through the posterior fossa in the spinal canal. The patient in question stated that she had experienced intense headaches before diagnosis, often located in the occiput or neck pain. However, the characteristic symptoms of this malformation, such as motor deficiency, weakness, sensory disturbances, unstable gait (ataxia), and pain, are present in the patient in question.

Some complementary exams to be requested are computed tomography of the skull and thorax, abdominal ultrasound, magnetic resonance imaging of the brain, echocardiogram with color Doppler, obstetric ultrasound, computed tomography of the brain segment, thyroid ultrasound, quantitative EEG with brain mapping, electroencephalogram, and measurement of cerebrospinal fluid (CSF) pressure. In this way, this study shows that a diagnosed entity is rare and important to medical knowledge[6].

Conclusion

It is concluded that the patient suffering from type I Chiari malformation is leading to obstructive hydrocephalus and can be treated with multidisciplinary monitoring by doctors and physiotherapists. Medical treatment can be through surgical interventions along with therapeutic interventions, the surgical treatment will be related to the patient's two conditions, that is, decompression of the posterior fossa will be carried out, so that there is decompression of the brain stem and the high spinal cord, bringing, therefore, improves the symptoms caused by the disease. Regarding obstructive hydrocephalus, surgical or therapeutic treatment can be carried out, but the best choice must be made depending on the evolution of the Chiari I malformation, if it does not evolve again, definitive treatment must be carried out, that is, a surgical intervention, using the “shunt” valve technique. Now, if it evolves again, surgical, and therapeutic treatment must be carried out, the surgical treatment will be the same if the swelling does not evolve further. The therapeutic is done as follows, acetazolamide must be used, and furosemides can reduce CSF production by 50 – 60%, as carbonic anhydrase reducers. The recommended doses for acetazolamide are between 50 and 150 mg/kg/day and for furosemide it is 1 mg/kg/day.

In addition to the limited effect on the control of hydrocephalus and intracranial hypertension, side effects such as metabolic acidosis, demyelination, and nephrocalcinosis were observed. Osmotic diuretics, such as Isosorbide, Mannitol, Urea, and Glycerol, work by decreasing the water content of the brain. They can be used as a provisional measure in communicating hydrocephalus, but they are ineffective in bulky hydrocephalus due to parenchymal reduction. Among the most important side effects were the so-called rebound effect, hypernatremia, and dehydration. Corticosteroids such as dexamethasone and methylprednisolone have been indicated to stimulate CSF absorption, reducing the inflammatory response. Heparin and hyaluronidase have been used to unblock arachnoid granulations.

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