

IMPACTED CISTERNA MAGNA WITHOUT SYRINGOMYELIA ASSOCIATED WITH SPASTIC PARAPARESIS

Case report

José Alberto Gonçalves da Silva¹, Maria do Desterro Leiros da Costa², Maurus Marques de Almeida Holanda¹, Luiz Ricardo Santiago Melo¹, Antônio Fernandes Almeida de Araújo¹, André Pachelli Bezerra Viana³

ABSTRACT - We report on a 49 year old man with impacted cisterna magna without the presence of syringomyelia (SM). The clinical picture was characterized by spastic paraparesis. Magnetic resonance imaging depicted a cisterna magna filled by the cerebellar tonsils. Six months after osteodural-neural decompression of the posterior fossa there was resolution of neurological symptoms and signs with the exception of hyperactive patellar and Achilles reflexes.

KEY WORDS: tight cisterna magna, impacted cisterna magna, Chiari malformation, posterior fossa decompression, spastic paraparesis.

Cisterna magna impactada sem siringomielia associada com paraparesia espástica: relato de caso

RESUMO - Hans Chiari (1891, 1895) descreveu 4 tipos de anomalias cerebelares que, posteriormente, foram denominadas de malformação de Chiari (MC). Iskandar et al. (1998) relataram 5 casos de hidrosiringomielia (SM) sem herniação rombencefálica, nos quais as tonsilas cerebelares preenchiam a cisterna magna. Todos os casos melhoraram após a descompressão da fossa posterior com redução do tamanho da cavidade siringomiélica. Relatamos o caso de paciente de 49 anos com cisterna magna impactada sem a presença de siringo-hidromielia (SM). O quadro clínico caracterizava-se por paraparesia espástica. A ressonância magnética evidenciou a cisterna magna preenchida pelas tonsilas cerebelares. Seis meses após a descompressão osteodural-neural da fossa posterior, houve resolução dos sintomas e sinais neurológicos, com exceção da hiperatividade dos reflexos patelares e aquileus.

PALAVRAS-CHAVE: cisterna magna impactada, malformação de Chiari, descompressão da fossa posterior, paraparesia espástica.

Hans Chiari^{1,2} described four types of cerebellar anomalies (CM). Type I characterized by downward displacement of the cerebellar tonsils and the medial portions of the inferior cerebellar lobes which accompanied the medulla into the cervical spinal canal. Type II showed downward displacement of portions of the cerebellum (1891), and portions of the inferior vermis (1895), pons, medulla oblonga and, at least, part of lengthened fourth ventricle, which reached the disc C 4-C 5, into the enlarged cervical spinal canal. In type III, the hydrocephalic cerebellum, pons and medulla oblonga were inside a cervical meningo-

cele (hydroencephalocèles cerebellaris cervicalis), through a spina bifida of the first three cervical vertebrae. In type IV, there was hipoplasia of the cerebellum without herniation of cerebellar structures into the spinal canal. Iskandar et al.³ (1998) related five cases of syringohidromyelia (SM), in which the cisterna magna was filled by the cerebellar tonsils. After decompression of the posterior fossa, there was clinical improvement, as well as marked reduction in the size of the syrinx in all cases. The authors admitted that this dramatic response to decompression indicates that this entity has a Chiari-like pathophysiology.

Neurosurgical Unit Hospital Unimed, João Pessoa PB, Brazil: ¹Neurosurgeon; ²Neurologist; ³Neuroanesthesist.

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Dr. José Alberto Gonçalves da Silva - Avenida Minas Gerais 1150 - 58030-092 João Pessoa PB - Brasil.

The present publication is based on the rareness of the clinical picture characterized by spastic paraparesis, hypopallesthesia of the lower limbs and sexual impotency associated with impacted cisterna magna without SM. We did not find a similar report in the consulted literature (Medline and Lilacs).

CASE

A 49-year old male with a five-year history, of progressive loss of muscular strength of the lower limbs causing severe gait difficulty, sexual impotency and paraesthesia on the feet, occasionally ascending up to the knees. Neurological exam showed spastic paraparesis, marked hyperactive patellar and Achilles reflexes, bilateral Hoffmann

and Babinski signs, inexhaustible knee and ankle clonus associated with diminished pallesthesia of the lower limbs. The patient was infected by hepatitis C virus (HCV) and since 1999 has been receiving interferon therapy. Magnetic resonance imaging (MRI) depicted a cisterna magna filled by the cerebellar tonsils (Fig 1).

The osteodural-neural decompression of the posterior fossa using the Gonçalves da Silva technique, was carried out with the patient in sitting position. A large craniectomy of the posterior fossa was performed and after the dural opening in Y format the following abnormalities were observed (Fig 2): the cerebellar tonsils filled the cisterna magna without herniation into the spinal canal, the fourth ventricle and the foramen of Magendie were compressed by the cerebellar tonsils and we could identify only the left

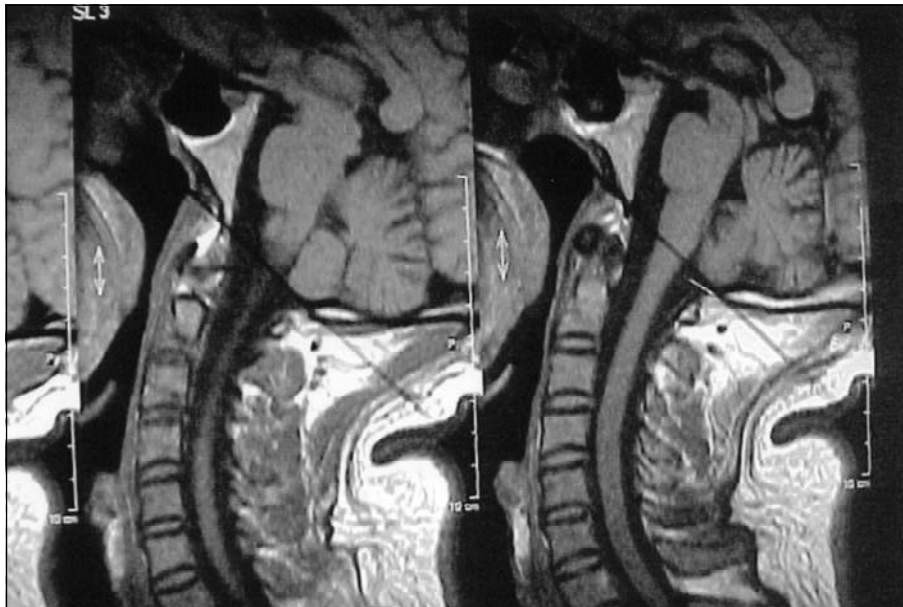


Fig 1. Impacted cisterna magna without SM.

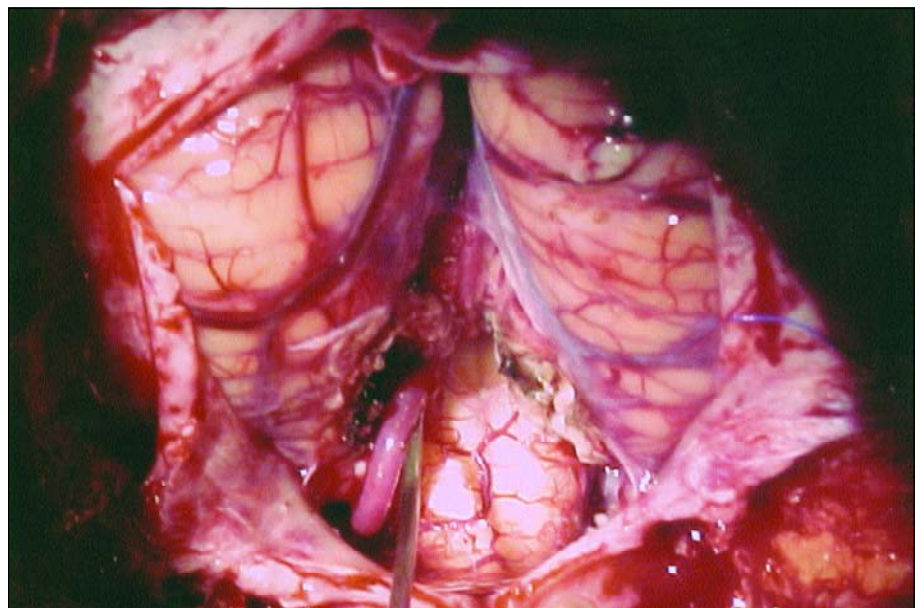


Fig 2. Tonsilectomy, large opening of the fourth ventricle and the Left PICA.



Fig 3. Postoperative MRI showing the new created cisterna magna.

posterior inferior cerebellar artery (PICA). After dissection of the arachnoid membrane, we performed intrapial aspiration of the cerebellar tonsils and made a large opening of the fourth ventricle, and sutured the residual pial sac upwards to the dura-mater in cranial lateral position.

The postoperative MRI depicted the large created cisterna magna and also that the cerebellar tonsils did not compress the fourth ventricle and the foramen of Magendie anymore (Fig 3). Six months after posterior fossa decompression the disappearance of neurological symptoms and signs was observed, with the exception of the hyperactive patellar and Achilles reflexes and amelioration of sexual impotency.

This study was approved by the appropriate Bioethics Research Committee.

DISCUSSION

Five cases of impacted cisterna magna without hindbrain herniation were related by Iskandar et al.³, nevertheless these cases were accompanied by SM. All cases improved after posterior fossa decompression with reduction in the size of the syrinx. The dramatic response to decompression indicates that this entity has a Chiari like pathophysiology. Kyoshima

et al.⁴ related four similar cases with improvement in symptoms and a reduction in syrinx size in three patients, and a reduction in ventricle size in two. The authors named "tight cisterna magna", the impacted cisterna magna by the cerebellar tonsils, and called the description according to Iskandar et al.³ "Chiari "0" malformation".

According to Williams⁵, the cerebellar tonsils herniation may compress brainstem structures and contribute to bulbar and a long tract dysfunction.

In a similar way the impactation of the cerebellar tonsils in the cisterna magna, without herniation into the cervical spinal canal causes disturbances in the CSF flow at the foramen magnum and can develop neurological symptomatology by compression of the brainstem. In the present case during the operation the fourth ventricle, brainstem and the foramen of Magendie were compressed by the impacted cerebellar tonsils. Probably this pathophysiology could explain the neurological symptoms and signs of this patient.

Spastic tetraparesis and paraparesis are frequently described in patients with basilar impression (BI) and/or CM and SM⁶⁻¹⁷, in cases of cervical spondilotic myelopathy¹⁸, constrictive arachnoiditis¹⁹, among others.

SM is absent in the present case, despite an evident obstruction to the CSF flow in the posterior fossa. This finding lead us to hypothesize whether in the future SM would develop, since obstruction to the CSF flow in the posterior fossa is considered the condition for its development.

Regarding the HCV, some authors described neurological complications like seizures, hemiparesis, hemianopsia, and urinary retention²⁰, vasculitic neuropathy²¹, neuropsychiatric symptoms associated with chronic HCV²². Highleyman²³ suggested that HCV coinfection adversely affects neuropsychological function in patients with HIV but does not seem to contribute to peripheral sensory neuropathy. The disappearance of the clinical symptomatology after the posterior fossa decompression excludes the HCV as part of the pathophysiology of this case.

More studies should be carried out regarding the impacted cisterna magna to elucidate its pathophysiology and the correct diagnosis for the surgical treatment. The study of CSF flow is a very important method for the diagnosis of the tight cisterna magna but unfortunately this exam is not at our disposal.

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