

BASILAR IMPRESSION ASSOCIATED WITH IMPACTED CISTERNA MAGNA, SPASTIC PARAPARESIS AND DISTRESS OF BALANCE

Case report

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ABSTRACT - We report on a 48 years-old man with basilar impression without syringohydromyelia, in which the cisterna magna was impacted by the cerebellar tonsils. Six months after posterior fossa decompression there was the disappearance of nuchal rigidity, vertigo, spastic paraparesis and improvement of balance. Nevertheless hyperreflexia and diminished pallesthesia of the lower limbs persisted.

KEY WORDS: basilar impression, Chiari malformation, syringomyelia, posterior fossa decompression, tight cisterna magna.

Impressão basilar associada com cisterna magna impactada, paraparesia espástica e distúrbios do equilíbrio: relato de caso

RESUMO - O presente relato descreve um homem de 48 anos com impressão basilar, sem siringo-hidromielia, no qual a cisterna magna se achava impactada pelas tonsilas cerebelares. O quadro clínico era caracterizado, especialmente, por paraparesia espástica. Seis meses após a decompressão da fossa posterior houve regressão da rigidez de nuca, vertigem, paraparesia espástica inicialmente observadas e melhora do equilíbrio. A hiperreflexia e a hipopalestesia persistiram nos membros inferiores.

PALAVRAS-CHAVE: impressão basilar, malformação de Chiari, siringomielia, decompressão da fossa posterior, cisterna magna impactada.

Hans Chiari^{1,2} (1891, 1895) described four types of cerebellar anomalies. Type I was 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. The type II showed downward displacement of portions of the cerebellum (1891), and portions of the inferior vermis (1895), pons, medulla oblonga and, at least, a 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 were inside a cervical meningocele (hydroencephalocèles cerebellaris cervicalis), through a spina bifida of the first three cervical vertebrae. In type IV, there was hypoplasia of the cerebellum without herniation of cere-

bellar structures into the spinal canal. Iskandar et al.³ (1998) related five cases of syringohydromyelia (SM), in which the cisterna magna was filled by the cerebellar tonsils. In all five cases there was clinical improvement after decompression of the posterior fossa and a marked reduction in the size of syrinx. These authors admitted that this dramatic response to decompression indicates that this entity has a Chiari-like pathophysiology. On the other hand, 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. These authors named the impacted cisterna magna by the cerebellar tonsils "tight cisterna magna", and called the description according to Iskandar et al.³, "Chiari "0" malformation".

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The present case offers some resemblances with the pathophysiology, described by Iskandar et al.³. This paper is based on the rareness of the clinical picture, characterized by brevis collis, nuchal rigidity, vertigo, spastic paraparesis, diminished unsteady gait and diminished pallesthesia of the lower limbs.

CASE

A 48-year-old man presented, with a history of progressive loss of muscular strength of the lower limbs, daily vertigo and distress of balance. The neurological exam presented brevis collis, nuchal rigidity, distress of balance, spastic paraparesis, marked hyperactive patellar and Achilles reflexes and diminished pallesthesia of the lower limbs. The X rays of the skull showed basilar impression (BI) and a normal anatomic configuration of the foramen magnum. The magnetic resonance imaging (MRI) depicted the presence of BI, absence of the cisterna magna and the brain stem seemed not to be compressed by the odontoid process (Fig 1).

The indication for the following surgical treatment is concerned to the impacted cisterna magna. During the osteodural-neural decompression of the posterior fossa, carried out with the patient in sitting position^{5,6}, we observed the following abnormalities: pulseless dura-mater, diffuse arachnoiditis, compression of the fourth ventricle and the foramen of Magendie by the cerebellar tonsils which filled the cisterna magna and migrated partially downwards into the cervical spinal canal (Fig 2). The cerebellar tonsils were yellowish, lightly atrophic and presented some arachnoid adhesions to neighboring structures. We performed intrapial aspiration of the cerebellar tonsils and the residual pial sac was sutured upwards to the dura-mater in cranial lateral position (Fig 3). Six months after posterior fossa decompression, we observed disappearance of nuchal rigidity, vertigo, spastic paraparesis and amelioration of the balance distress, while the signs of pyramidal liberation and the diminished pallesthesia of the lower limbs remained unchanged. The postoperative MRI revealed the created cisterna magna (Fig 4).



Fig 1. Impacted cisterna magna and BI.

ration of the balance distress, while the signs of pyramidal liberation and the diminished pallesthesia of the lower limbs remained unchanged. The postoperative MRI revealed the created cisterna magna (Fig 4).

This study was approved by the appropriate Bioethics Research Committee.

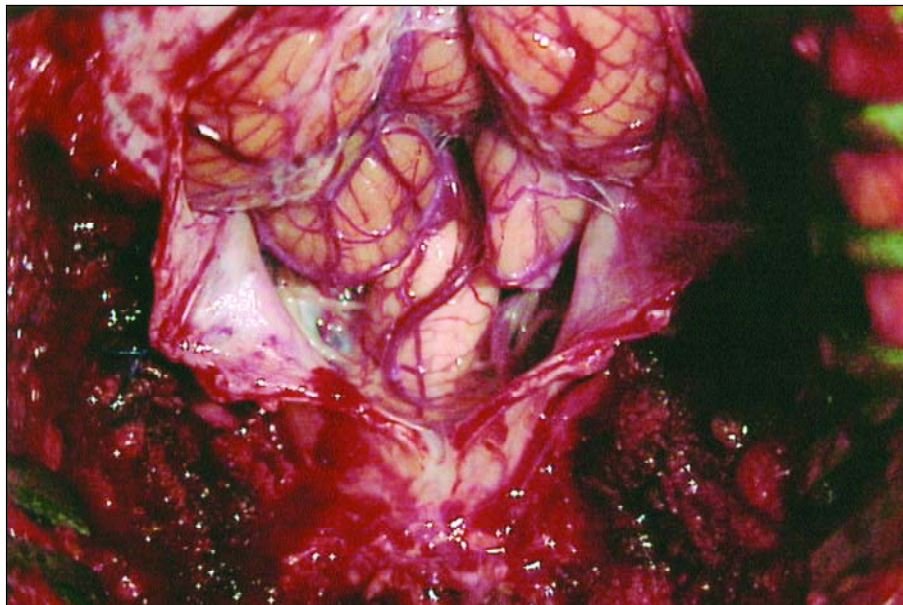


Fig 2. Partially downwards migration of the cerebellar tonsils during operation in sitting position.

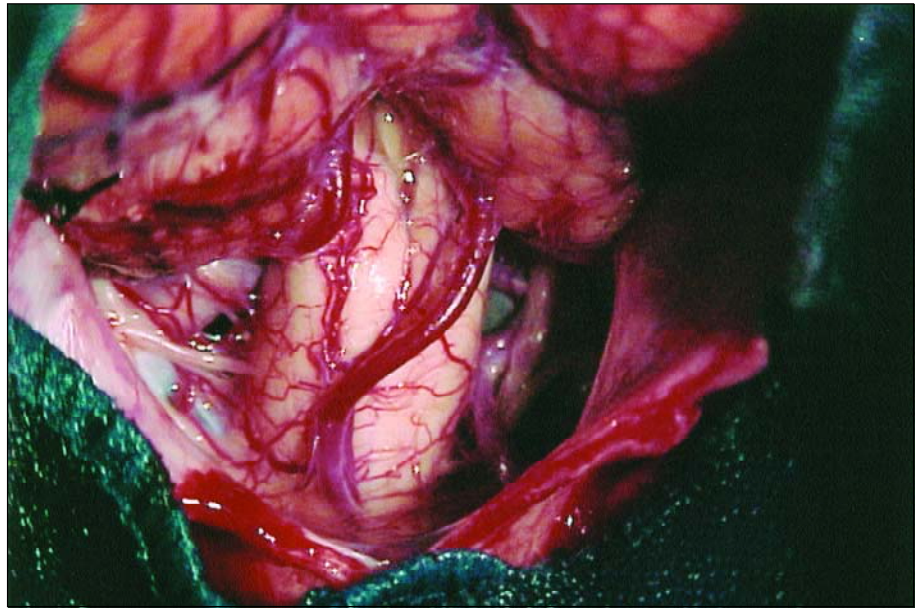


Fig 3. Tonsilectomy, large opening of the fourth ventricle and fixation of the residual pial sac to the lateral dura-mater in cranial lateral position.



Fig 4. Postoperative MRI showing the new created cisterna magna.

DISCUSSION

In the present report we describe the association of BI and impacted cisterna magna. The X rays showed the presence of BI and a normal anatomic configuration of the foramen magnum, whose posterior

border do not penetrate into the posterior fossa. The preoperative MRI depicted an impacted cisterna magna, without herniation into the cervical spinal canal, without SM. The brainstem seemed not to be compressed by the odontoid process. However, during the operation performed with the patient in sitting position, we noticed small herniation of the cerebellar tonsils below the foramen magnum. This observation was also described by Iskandar et al.³ and Gonçalves da Silva et al.⁷, in which the cerebellar tonsils were located, during the operation, below the foramen magnum. Possibly the cerebellar tonsils have a tendency to migrate downwards on the ortostatic position. In the future, with the introduction of MRI carried out with the patient in upright position, we probably shall be able to detect a difference of the cerebellar tonsils topography in both the dorsal and the ortostatic position.

The impactation of the cerebellar tonsils in the cisterna magna, without herniation into the cervical spinal canal, also causes disturbances of the cerebral spinal fluid (CSF) flow to the foramen magnum, according to Iskandar et al.³, and can provoke neurological symptomatology by compression of the brainstem and the cerebellar pathways.

Spastic tetraparesis and paraparesis are frequently described in patients with BI and/or Chiari malformations (CM), in which distress of balance is observed in cases of CM⁸⁻¹⁶.

The pathophysiology of this case could have been developed either by BI or the impacted cisterna magna. However, the BI does not show signs of com-

pression of the brainstem and the posterior border of the foramen magnum does not penetrate into the posterior fossa. Possibly the brevis collis and the nuchal rigidity were caused by the BI, while the impacted cisterna magna developed other signs like vertigo, distress of balance, spastic paraparesis and diminished pallesthesia of the lower limbs.

More studies should be carried out regarding impacted cisterna magna, isolated or associated with other pathologies, with the purpose of defining correctly this diagnosis and the subsequent posterior fossa decompression. 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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