

# HERNIATION OF THE CEREBELLAR TONSILS AFTER SUPRASELLAR ARACHNOID CYST SHUNT

## Case report

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**ABSTRACT** - It is known that the caudal dislocation of the cerebellar tonsils may occur associated with clinical conditions such as an intracranial mass lesion or Chiari I and II malformation. It may also be acquired after repeated lumbar punctures or lumboperitoneostomy. The occurrence of cerebellar herniation after derivation of intracranial arachnoid cyst is extremely rare, and there are only three cases reported in the medical literature. We present the case of a 9-year-old boy with precocious puberty and suprasellar arachnoid cyst who developed a symptomatic herniation of the cerebellar tonsils three years after a cystoperitoneostomy. The patient underwent a suboccipital craniectomy with duraplasty and partial tonsilectomy, showing afterwards, remission of the symptoms. We discussed the pathogenesis suggested in the literature.

**KEY WORDS:** Arnold Chiari malformation, arachnoid cyst.

### **Herniação das tonsilas cerebelares após *shunt* de cisto aracnóide supra-selar: relato de caso**

**RESUMO** - Sabe-se que o deslocamento caudal das tonsilas cerebelares pode ocorrer em associação com condições clínicas tais como: lesão expansiva intracraniana ou malformação de Chiari I e II. Pode ainda ser adquirido após repetidas punções lombares ou lumboperitoniotomia. A ocorrência de herniação cerebelar após derivação de cisto aracnóide intracraniano é evento extremamente raro, existindo apenas três casos relatados na literatura médica. O caso de menino de 9 anos de idade, com puberdade precoce e cisto aracnóide supra-selar, que desenvolveu herniação sintomática das tonsilas cerebelares três anos após cistoperitoniotomia. O paciente foi submetido a craniectomia suboccipital com plástica dural e tonsilectomia parcial, apresentando remissão dos sintomas. Discutimos a patogênese sugerida na literatura.

**PALAVRAS-CHAVE:** malformação de Arnold-Chiari adquirida, cistos aracnóides.

The term "Acquired Chiari I" has been used to describe the herniation of the cerebellar tonsils secondary to intracranial hypotension due to multiple lumbar punctures or a lumboperitoneal derivation. Neuroimage and clinical observations have demonstrated the possibility of later herniation of the structures of the posterior fossa in patients who did not have such alteration at birth. The mechanism involved in the rare cases of acquired Chiari I - specially in those secondary to a supratentorial shunt - is still unknown. Cephalocranial disproportion has been implicated<sup>1</sup>.

### **CASE**

A 12 years old boy, caucasian, was admitted with inca-

pacitating nape pain. The patient had a previous history of suprasellar arachnoid cyst and precocious puberty (Fig 1), having undergone derivation of arachnoid cyst to the peritoneal cavity (cyst-ventricle-peritoneostomy) in January 2002. The neurological examination did not show any alterations. Investigation with cranial magnetic resonance (MR) showed acquired herniation of the cerebellar tonsils (Fig 2) and upper herniation of the cerebellar culmen, with an increase volume of the neural structures of the posterior fossa.

The patient underwent decompressive surgery with suboccipital craniectomy, with removal of the posterior arch of C1, bipolar coagulation and partial aspiration of the cerebellar tonsil and duraplasty with periosteum. There was immediate improvement of the symptoms, without any post-operative complications. Control MR showed an ade-

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Fig 1. Suprasellar arachnoid cyst (large arrow). Notice the size of prepontine cystern (two small arrows), the size of ambient cystern (head of arrow), and the relationship of the tonsils and foramen magnum.



Fig 2. After the cystoperitoneal shunt (large arrow), there was a increase in the volume of contents of the posterior fossa, with diminution of prepontine (double arrows) and ambient cysterns, and tonsillar herniation above the foramen magnum. Notice the upper herniation of the culmen cerebellar (head of arrow), and its proximity with the splenium of corpus callosum.

quate decompression of the posterior fossa and the regression of the tonsillar herniation (Fig 3).

The parents of the patient agree with the report of the case.

**DISCUSSION**

It is known that the caudal dislocation of the cerebellar tonsils may occur associated with clinical conditions, such as an intracranial mass lesion or Chiari I and II malformation. It may also be acquired after repeated lumbar punctures or lumboperitoneostomy. The occurrence of cerebellar herniation after derivation of intracranial arachnoid cyst is extremely rare, having only three cases been reported in the medical literature, upon Medline and LILACS<sup>2-4</sup> database consultation.

Hoffman and Tucker<sup>1</sup> have proposed that a temporary interruption in the growth of the cranium subsequent to a collapse of the ventricular system leads to a cephalocranial disproportion, which would help the herniation of the cerebellar tonsils through the foramen magnum.

Di Rocco and Velardi<sup>2</sup> have described a case of symptomatic Chiari I, years after derivation of an arachnoid cyst to the peritoneal cavity. A parietal bilateral decompressive supratentorial craniectomy was performed, resulting in improvement of the symptoms. The authors emphasize that the secondary cephalocranial disproportion is a relevant factor



Fig 3. Post-operative control, showing the posterior fossa craniectomy and duraplasty (three arrows), with regression of tonsillar herniation. Note the persistence of the herniation of culmen cerebellar (head of arrow).

in the acquisition of a cerebellar herniation. The precocious craniosynostosis due to the derivation, that is, the interruption in the growth of the skull, associated with the continuous growth of the nervous tissue, would be the factors responsible for cerebellar tonsillar herniation.

Lazareff et al.<sup>3</sup> described a case of an 8-year-old boy whose cerebellar tonsil herniation occurred just after the collocation of a cystoperitoneal shunt. The authors emphasize the role of structural posterior fossa abnormalities on the development of the herniation of the cerebellar tonsils, and point out that only those patients who have a congenial predisposition will develop the acquired Chiari I condition.

Hassounah and Rahm<sup>4</sup> reported a case of herniation of the cerebellar tonsils secondary to the shunt of an intracranial arachnoid cyst. They proposed the occurrence of a difference of pressure between the medullar and cranial compartments after the shunt. Another mechanism suggested was the reexpansion of the neural tissue inside the posterior fossa after the drainage of the cyst. The cephalocranial disproportion proposed by Hoffman and Tucker may as well apply to this case.

Jakob<sup>5</sup> makes a morphological and biochemical description of the brain, pointing out that the cerebellum would be the most "hydric" portion of the central nervous system, that is, the most easily hydrated part, which could explain the increase volume of the content of the posterior fossa upon shunt of the

cyst, which supports the report of Hassounah and Rahm<sup>4</sup>.

In conclusion, the herniation of the cerebellar tonsils after supratentorial shunt is extremely rare. The pathogenesis suggested in the literature, in the cases with difference of pressure between the spinal and cranial compartments and cephalocranial disproportion due to the precocious closure of cranial sutures, does not apply in this case, since the patient already had closed sutures at the time of the cystoperitoneal shunt. The increase volume of the contents of the posterior fossa, since the cerebellum is the most easily hydrated portion of the central nervous system<sup>5</sup>, may have a role in the herniation of the tonsils and cerebellar culmen in the case discussed here.

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