Improvement of Hydrocephalus in Patients with Chiari Malformation After Posterior Fossa Decompression. Case Report

Melhora da Hidrocefalia em Paciente com Malformação de Chiari Após Descompressão de Fossa Posterior. Relato de Caso

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ABSTRACT

Introduction. The association between hydrocephalus and Chiari malformation (CM) has not been described frequently. Ventricular dilation affects 7% to 10% of patients with CM, but the ideal choice of surgical treatment is controversial. We report a case of a patient with Chiari malformation and hydrocephalus with improvement in clinical symptoms and ventricular dilatation. Case Description. A 19-year-old male complaining of frontal headache when coughing, laughing and during valsalva maneuvers, associated with dizziness for 2 months. Magnetic resonance (MR) showed hydrocephalus and small posterior fossa with overcrowding of contents, characterizing Chiari malformation type I, with cerebellar tonsils protruding through magnum foramen. Patient underwent surgery with posterior fossa decompression in a semi-sitting position and removal of the arc C1. After 3 months of follow-up, headache disappeared becoming asymptomatic. Control MR showed improvement of hydrocephalus with restoration of the cisterna magna and CSF flow. Discussion. Hydrocephalus has been related to CM for a long time. In our case, we performed treatment with intradural and intra-arachnoidal approach with bilateral tonsillectomy without placing ventricular shunt. The cisterna magna was “recreated”. There was improvement of hydrocephalus with decreased Evans ratio index and symptoms disappearance. Although there is no other studies addressing such matter, in this case, the improvement suggests that the CSF compression at the foramen magnum was the cause of associated hydrocephalus with Chiari malformation.

Key words: Chiari malformation; Hydrocephalus; Posterior fossa decompression

RESUMO

Introdução. A associação entre hidrocefalia e malformação de Chiari (CM) não foi descrita com frequência. A dilatação ventricular afeta 7% a 10% dos pacientes com CM, mas a escolha ideal do tratamento cirúrgico é controversa. Relatamos um caso de um paciente com MC e hidrocefalia com melhora nos sintomas clínicos e dilatação ventricular após tratamento.Descrição do caso. Homem de 19 anos queixando-se de cefaleia frontal durante tosse, riso e manobra de Valsalva, associado a tonturas por 2 meses. A ressonância magnética (RM) mostrou hidrocefalia e fossa posterior pequena, caracterizando a malformação de Chiari tipo I, com tonsilas cerebelares protraindo através do forame magno. Paciente submetido a cirurgia com descompressão da fossa posterior em posição semi-sentada e remoção do arco de C1. Após 3 meses de acompanhamento houve desaparecimento de cefaleia, tornando-se assintomático. A RM controle mostrou melhora da hidrocefalia com restauração da cisterna magna e fluxo de LCR. Discussão. A hidrocefalia tem sido relacionada a CM há muito tempo. No nosso caso, realizamos tratamento com abordagem intradural e intra-aracnoidal com amigdalectomia bilateral sem colocação de shunt ventricular. A cisterna magna foi recriada. Houve melhora da hidrocefalia com diminuição do índice de Evans e desaparecimento dos sintomas. Embora não haja outros estudos abordando essa questão, neste caso, a melhora sugere que a compressão do LCR no forame magno foi a causa da hidrocefalia associada à malformação de Chiari.

Palavras-chave: Malformação de Chiari; Hidrocefalia; Descompressão da fossa posterior

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INTRODUCTION

The association between hydrocephalus and Chiari malformation (CM) has not been described frequently. Ventricular dilation affects 7% to 10% of patients with CM but the ideal choice of surgical treatment is controversial1-7.

The compression of cerebrospinal fluid (CSF) outflow tract hinders CSF circulation and predisposes hydrocephalus3. However, the presence of hydrocephalus and CM has occurred in a minority of cases and thus the need for treatment of both CM and hydrocephalus is not fully understood and defined. Several authors propose the treatment of hydrocephalus initially sometimes not requiring subsequent posterior fossa decompression (PFD). However, others state that PFD should be the initial approach, maybe avoiding a shunt2,5.

We report a case of a patient with Chiari malformation and hydrocephalus with improvement in clinical symptoms and ventricular dilatation.

CASE DESCRIPTION

A 19-year-old male complaining of frontal headache when coughing, laughing and valsalva maneuvers, associated with dizziness for 2 months searched for evaluation in the São Luiz hospital, São Paulo, Brazil. The clinical picture was longstanding, since childhood, with improvement and worsening periods which has intensified in the last two months. He denied suboccipital pain. Neurological examination was normal.

Magnetic resonance (MR) showed hydrocephalus and small posterior fossa with overcrowding of contents, characterizing Chiari malformation type I with cerebellar tonsils protruding through magnum foramen. Preoperative Evans ratio was 0.34 (Figure 1).

Patient underwent surgery with posterior fossa decompression in a semi-sitting position and removal of the arc C1. An intradural and intra-arachnoidal technique was applied (including bilateral tonsillectomy and duraplasty). Postoperative period was uneventful and patient was discharged after 3 days.

After 3 months of follow up, there was disappearance of headache, becoming asymptomatic. Control MR showed improvement of hydrocephalus with restoration of the cisterna magna and CSF flow. Postoperative Evans ratio is now 0.31 (Figure 1).

Figure 1. Above, preoperative MRI revealing Chiari Malformation and hydrocephalus, Evans ratio of 0.34. Below, postoperative images showing posterior fossa decompression, partial resection of cerebellar tonsils, creation of a new cisterna magna and decrease in Evans ratio (0.31) suggesting improvement of CSF circulation after surgery.

DISCUSSION

The relationship between hydrocephalus and CM is not elucidated. The pathogenesis of this association is complex. Supratentorial hypertensive hydrocephalus may also cause tonsillar herniation, as well as any supra or infratentorial hypertension exerting pressure from above to the cerebellum1-7. Hydrocephalus has been related to CM for a long time3. However, the distinguishing factor is that in primary CM, the tonsil herniation often occurs in situations where the volumetric capacity of the posterior fossa (PF) is small and herniation of the tonsils occurs without intracranial hypertension. Nevertheless, CM can lead or be associated to hydrocephalus due to a disorder of CSF flow in the craniocervical junction3.

There are few studies describing the association between the two entities. Sahuquillo et al. described the technique of reconstruction of the posterior fossa and reported five patients with hydrocephalus and CM. Of these, only one received shunt before PFD and other needed shunt early after PFD6. Deng et al. described 38 adult patients who received PFD as
treatment for CM with hydrocephalus. Postoperative magnetic resonance (MR) images showed relief of cervicomedullary compression and recreation of the cisterna magna. No significant change in ventricular size was observed after surgery, but symptoms improved in 33 patients (86.8%). They stated PFD with duraplasty as an effective and safe treatment for CM with ventricular dilatation and proposed that treatment of ventricular dilatation is unnecessary before PFD.

In our case, MR shows a small occipital bone and subsequent overcrowding of posterior fossa. We performed treatment with intradural and intra-arachnoidal approach with bilateral tonsillectomy without placing ventricular shunt. The cisterna magna was “recreated”. There was improvement of hydrocephalus with decreased Evans ratio index and symptoms disappearance. Release of the compression of the foramen of Magendie and Luschka and restoration of CSF flow may have contributed to the treatment of hydrocephalus.

The genesis of hydrocephalus in cases of CM is complex and probably multifactorial. Relief of hydrocephalus in our patient suggests that the overcrowding of the posterior fossa structures contributed to the genesis of hydrocephalus. PFD was sufficient to treat CM and hydrocephalus.

Although there are no other studies addressing such matter, in this case, the improvement suggests that, the CSF compression at the foramen magnum was the cause of associated hydrocephalus with Chiari malformation.

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