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SYRINGOHYDROMYELIA ASSOCIATED TO THERAPEUTIC PROCEDURES FOR SEVERE FORMS OF NEUROCYSTICERCOSIS

Case report

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ABSTRACT - Syringohydromyelia is defined as a longitudinal dilatation of the central canal of the spinal cord with accumulated cerebrospinal fluid. This condition may cause neurologic deficits when the cavity enlarges and compresses the spinal cord. We present the case of a 33 years-old female with progressive paraparesis caused by syringohydromyelia. This patient underwent previously multiple clinical and surgical treatments for severe form of neurocysticercosis. Surgical decompression of the posterior fossa and syringostomy resolved the neurologic symptoms. The possibility of syringohydromyelia should be considered in the case of patients who have previously undergone surgical and clinical treatment for severe form of neurocysticercosis.

KEY WORDS: syringohydromyelia, neurocysticercosis, surgical treatment.

Hidrossiringomielia associada ao tratamento de formas severas de neurocisticercose: relato de caso

RESUMO - A hidrossiringomielia é definida como uma dilatação longitudinal do canal central da medula espinhal cujo conteúdo é semelhante ao líquido cefalorraquidiano e que freqüentemente produz comprometimento neurológico. Apresentamos o caso de uma paciente de 33 anos, submetida a vários tratamentos clínicos e cirúrgicos para neurocisticercose severa, que tardiamente apresentou quadro de paraparesia progressiva, quando foi então, detectada uma hidrosiringomielia. O tratamento cirúrgico obteve resolução do quadro neurológico e considerável diminuição da cavidade hidrosiringomiélica. A possibilidade do desenvolvimento de hidrossiringomielia nas formas severas de neurocisticercose submetidas a vários tipos de tratamento deve ser suspeitada e investigada.

PALAVRAS-CHAVE: hidrossiringomielia, neurocisticercose, tratamento cirúrgico.

Over the last decades, neurocysticercosis (NC) has become the most common parasite affecting of the central nervous system and it is an endemic disease in Asia, Africa and Latin America[1,2]. This parasite occurs in the human due to a deviation in the life cycle of the *Taenia solium* in the pig, its usual intermediary host. There are two well defined larval forms: Cysticercus cellulosae *(cysts with a diameter of 1 to 20 mm containing a scolex) and Cysticercus racemosus* (cysts of variable aspects with a diameter of 40 to 120). The first type occurs within the nervous parenchyma and the second in the subarachnoid cisterns and ventricles. There is a third form (intermediary) presenting multiple membranous formations with a scolex. The clinical and laboratory condition of NC is quite polymorphic. Diagnosis is based on clinical observations, cerebrospinal fluid (CSF) examinations and neuroimaging findings, mainly the computerized tomography (CT) and magnetic resonance (MRI)[2].

Syringohydromyelia is the dilation of the central canal of the spinal cord due to accumulation of a fluid similar to CSF. These cavities differ from the syringomyelia because they are lined with the ependyma and are almost always associated with the hydrocephaly[3]. However, the term syringomyelia includes the various kinds of posttraumatic cysts that contain CSF, cysts associated with most of the tumors of the spinal medulla and even post-infected medullar cysts[4]. Communication of the

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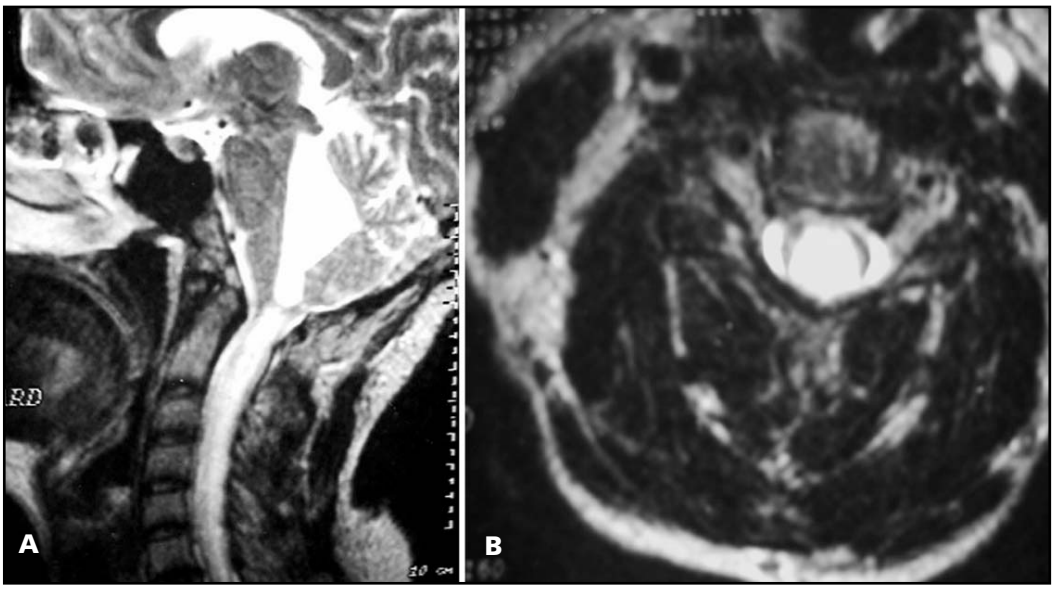

Fig 1. T2-weigted MRI ((A) sagital view, and (B) axial view) reveals a large cervical syringohydromyelic cavity.

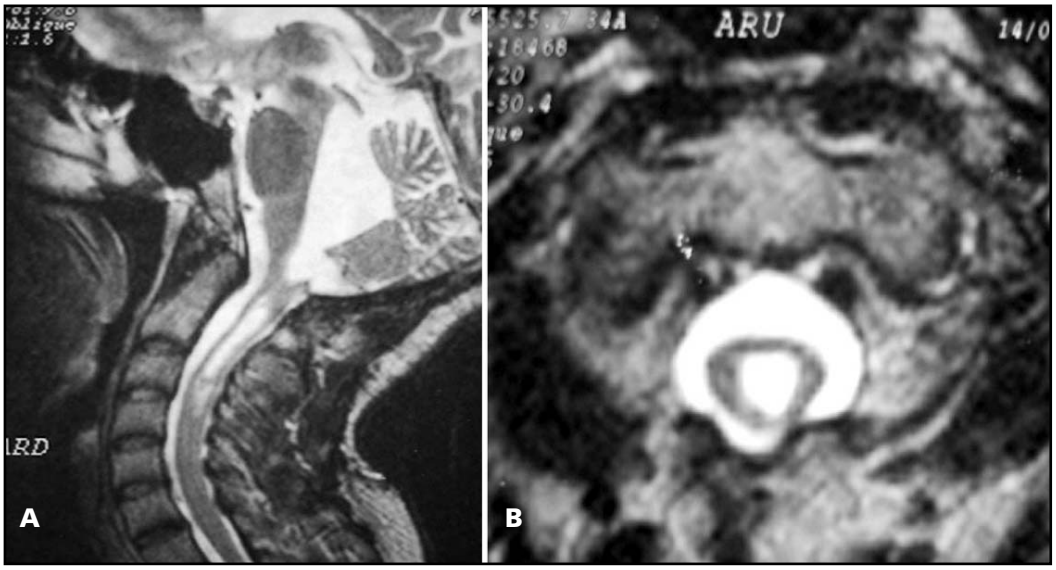

Fig 2. A post-operative (A) sagittal and (B) axial MRI (T2 WI) showing reduction of cervical syringohydromyelic cavity.

cavity with the IV ventricle is not common in syringomyelia occurring in only in only 10% of the cases, but in syringohydromyelia it is very common. Overall, all longitudinal medullar cavities are included in the term syringomyelia. Generally two types are identified: syringobulbia and syringomyelia. Syringobulbia occurs in the brain stem (medulla oblongata and cervical spinal cord) and has several causes: bone anomalies at the cranio-vertebral junction; arachnoidal adherences and posterior fossa anomalies as the Arnold-Chiari syndrome. Syringomyelia is a longitudinal cavity of the spinal cord due to posttraumatic, post-surgical and post-infectious arachanoid adherences, spinal cord medullar tumors and other pathologies associated with spinal dysraphism[4-6]. Syringomyelia is associated with blockage of the CSF circulation at the site or at a distance of the cavity. This fact implies that obstruction of the subarachanoid space impedes the spontaneous equilibration of the CSF pressure. It may occur even under physiological circumstances as pulsation of the blood vessels in the choroid plexus, repetitive physical efforts and coughing episodes. These situations increase the perimedullar CSF

pressure provoking CSF flow into the spinal cord through Virchow-Robin space.

A patient presenting syringohydromyelia after multiple treatments for severe form of neurocysticercose is reported. A similar case could not be found in the literature.

CASE

A 33-year old white female started in June 1998 to present symptoms of increased intracranial pressure (ICP). Hydrocephalus was diagnosed and ventricle-peritoneal shunts (VPS) was inserted at another department. In September 1998 a cysticercotic cyst was removed from the left temporal region and treatment with albendazole was carried out. On January 7, 1999 she was admitted to our clinic with symptoms of cysticercotic meningitis. The diagnosis was confirmed by CSF examination. Her clinical condition and CSF improved (ELISA test for cysticercosis was negative) after new treatment with albendazole. This therapy was repeated in February 1999 and February 2000. In March 2000, the patient complained frequent dizziness, nausea and occasional vomiting. A MRI-examination showed diffuse arachnoiditis and a cyst in the IV ventricle. On March 13, 2000, she underwent a craniectomy of the posterior fossa to remove the IV ventricle cyst and open the diffuse arachnoiditis. The clinical condition improved after operation and the patient became asymptomatic. One month after surgery she developed an infection of the distal cateter of the VPS with an abscess of the abdominal wall and meningitis. The infection (caused by oxacillin -sensitive *Staphylococcus aureus*) was adequately treated with antibiotics. The entire VPS was removed and an external ventricular drainage was inserted. A low pressure shunt was implanted three weeks later. The patient showed a significant improvement. A control MRI was performed in July 2001, and revealed dilatation of the IV ventricle due to obstruction of the inferior aqueduct orifice and the lateral (Luschka) and median foramens (Magendie) of the IV ventricle. The magnum foramen was obliterated by the cerebellar tonsils and obex. Cervical syringohydromyelia and suffusion in the medullar parenchyma were also observed. The patient remained asymptomatic, and no surgical treatment was proposed.

In December 2001, she developed progressive spastic paraparesis and a MRI revealed increase of the hydrosyringomyelia (Fig 1). On December 3, 2001 a new posterior fossa exposure, widening the previous craniectomy and communicating the hydrosyringomyelia with the subarachnoid space, was performed. Postoperatively the patient presented remission of paraparesis. She presented no deficit at the last neurological control examination (July 2003). The postoperative MRI demonstrated the reestablishment of normal CSF dynamics in the posterior fossa with reduction of the cervical syringohydromyelic cavity (Fig 2).

DISCUSSION

The clinical manifestations of NC are unspecific and varied. This polymorphism is due to individual differences in number, size and location of the lesions as well as the immunologic response of the host. Epilepsy is the most common presentation of NC (50-80%) and is usually the first or only clinical symptom of disease. Headaches as an isolated symptom occur in about 20% of the cases and intracranial hypertension in 15%. Meningitis is present in 8% and other clinical combinations are present in 2%[7,8].

Neurocysticercosis presenting cysts or calcification within the cerebral parenchyma are termed *Cysticercus cellulosae* and has the best prognosis with clinical treatment. *Cysticercus racemose* tends to be more severe and the parasite is generally lodged in the arachnoid cisterns or inside the ventricles. Occurrence of vasculits and hydrocephalus is frequent in this form of NC and clinical treatment is not always effective. Our patient presented this clinical condition.

Several researchers state that the presence of hydrocephaly qualify the condition as a severe form of NC. According to Torrealba et al.[9] and Agapejev et al.[2] the prognosis is worse when hydrocephalus is the first manifestation. Insertion of a VPS should be performed before cystical treatment, which should begin as soon as possible[2,8]. In our case the low pressure VPS, probably reduced the CSF flow in the cerebral aqueduct resulting in an isolated IV ventricle. The repeated clinical treatment, the direct approach to the IV ventricle and the ventriculitis affected the CSF circulation at the cranio-vertebral joint. Obstruction of the inferior orifice of the aqueduct and the ventricles exits, observed in the MRI, probably caused the CSF pulse wave at each systole. These waves directed towards the central canal through the obex, caused dilation and formation of the syringohydromyelic cavity, corroborating Gardner's theory[10]. The diagnosis of syringohydromyelia should be considered when patients treated for severe forms of NC present pyramidal signs and paraparesis. An isolated IV ventricle is usually easily identifiable in the CT and MRI[11], but the cervical segments of the spinal cord are not studied in these cases, impeding the diagnosis of syringohydromyelia. Several surgical techniques were described for treatment of syringomyelia[6]. The procedure used to reestablish CSF circulation in the posterior fossa of our patient was widening the craniectomy by resection of the posterior arch of the

atlas, opening the arachnoid adherences that blocked the IV ventricle, and create a communication between the syringomyelic cavity and the magnum cistern and the cervical perimedullar space. This procedure reestablished CSF circulation in the posterior fossa as demonstrated by the MRI (Fig 2).

In conclusion, in severe forms of NC with hydrocephaly, a VPS should be the first procedure. Clinical treatment with cysticidal drugs should be started as soon as possible. Surgery of the posterior fossa associated with clinical treatment in this form may cause severe inflammatory reaction with ventriculitis. Radiological investigation of the cervical region is required because syringohydromyelia may be diagnosed before these patients develop clinical symptoms. The possible development of syringohydromyelia should be considered in all patients with severe forms of neurocysticercosis.

REFERENCES

1. Spina França A, Livramento JA, Machado LR. Cysticercosis of central nervous system and cerebrospinal fluid: immunodiagnosis of 1573 patients in 63 years (1929-1992). Arq Neuropsiquiatr, 1993;51:16-20.
2. Agapejev S, Silva MD, Ueda AK. Severe forms of neurocysticercosis: treatment with albendazole. Arq Neuropsiquiatr 1996;54:82-93.
3. Wisoff JH, Epstein F. Management of hydromyelia. Neurosurgery 1989;25:262-271.
4. Sullivan LP, Stears JC, Ringel SP. Etiology of syringomyelia. Neurology 1989;39:308.
5. Arruda JAM, Costa CMC, Tella OI Jr. Avaliação de pacientes com siringomielia associada a malformação de Chiari: proposta de um protocolo multicêntrico. J Bras Neurocirurg 1999;10:93-96.
6. Costa V. O tratamento cirúrgico da siringomielia. Neurocirurgia Contemporânea Brasileira 2001; Vol 3/5.
7. Mareti MA. Neurocysticercosis: a study of 177 cases from São Jose do Rio Preto SP, Brazil. Arq Neuropsiquiatr 1999;57:1063.
8. Estañol B, Corona T, Abad P. A prognostic classification of cerebral cysticercosis: therapeutic implications. J Neurol Neurosurg Psychiatry 1986;49:1131-1134.
9. Torrealba G, Del Villar S, Tagle P, Arriagada P, Kase CS. Cysticercosis of the central nervous system: clinical and therapeutic considerations. J Neurol Neurosurg Psychiatry 1984;47:784-790.
10. Gardner WJ, Angel J. The mechanism of syringomyelia and its surgical correction. Clin Neurosurg 1959;6:131-140.
11. Colli BO, Pereira CU, Assirati JA, Machado HR. Isolated fourth ventricle in neurocysticercosis: pathophysiology, diagnosis and treatment. Surg Neurol 1993;39:305-310.