Une pachyméningite chez une fille âgée de 12 ans révélée par des céphalées et une confusion

Pachymeningitis revealed by acute confusion and headache in a twelve-year-old girl

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RESUME :

La pachyméningite est une entité rare chez l'enfant, avec des conséquences neurologiques potentiellement invalidantes. Elles correspondent à un épaississement inflammatoire chronique de la dure mère. Nous rapportons le cas d'une fille âgée de 12 ans qui a été hospitalisée pour céphalée aigue et une confusion. L'IRM cérébrale a montré une pachyméningite nodulaire disséminée. Les investigations à visée étiologique étaient normales. Après un recul de 12 mois, la patiente est restée asymptomatique.

Mots Clés : pachyméningite ; céphalée ; imagerie ; enfant.

ABSTRACT :

Pachymeningitis is a rare condition in children with potentially severe disabling neurological consequences. It's characterized by chronic inflammation causing thickening of the dura. We report here a 12 year-old-girl who presented with acute headache and confusion. Brain MRI showed diffuse nodular pachymeningitis. All etiological investigations were normal. After a follow up of 12 months, the patient remained asymptomatic without any neurological deficits.

Key words : pachymeningitis; headache; imaging; child.

INTRODUCTION :

Cranial pachymeningitis corresponds to chronic inflammatory thickening of the dura mater [1]; which can be caused by various infectious, autoimmune or malignant diseases. We report a case of nodular pachymeningitis, revealed by an acute headache and confusion in a 12 year old girl.

CASE REPORT :

A 12-year-old girl with a history of recurrent mouth ulcers was admitted for confusion, headache and vomiting. On examination, she had a temperature of 38.2°C. She was conscious and showed signs of intracranial hypertension without sensory-motor deficit or cranial nerve involvement and without meningeal syndrome. The fundus did not show papilledema. Laboratory investigations revealed no inflammatory syndrome. The computed tomography scan of the head was normal. Lumbar puncture showed pleiocytosis at 18 elements / mm3, normal sugar levels and high protein content at 8.3 g / l, the culture was negative. Brain MRI showed diffuse nodular pachymeningitis in the supra and infra-tentorial regions, particularly in the basal cisterns (Figure 1). This nodular aspect of pachymeningitis was in favor of either neuro-meningeal tuberculosis or sarcoidosis. The chest X-ray was normal and sputum and urine cultures were negative for tuberculosis. The lip biopsy did not show gigantic cell granuloma. The phosphocalcic balance was normal as well as the Angiotensin Converting Enzyme levels. The other causes of pachymeningitis

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were sought: aspergillus, syphilis and Lyme disease antibodies were negative, anti-nuclear antibodies, ANCA and anti-phospholipid antibodies were also negative. The HLA typing revealed B51. The thoraco-abdominal scan was without any abnormalities and the measurement of intracranial pressure was normal at 19 cm H2O. Recurrent aphthosis and HLA B51 typing were in favour of Behcet's disease. However the absence of other diagnostic criteria (no genital ulcers, no characteristic skin lesions, absence of uveitis and negative pathergia test) ruled out this diagnosis. Anti-tuberculosis treatment (quadritherapy) was initiated, later associated with oral corticosteroids (2 mg/kg /day which was tapered gradually within a month). Anti-tuberculosis drugs were stopped after 3 months when the culture of CSF for tuberculosis turned out to be negative. After four-year follow-up, our patient remained asymptomatic without any treatment.

DISCUSSION :

Pachymeningitis corresponds to chronic inflammatory thickening of the dura mater [1]. The clinical signs of pachymeningitis are variable and depend on the location of the lesions. Cranial pachymeningitis is usually revealed by headaches (40% of cases) and involvement of cranial nerves. Cerebellar ataxia and delirium can also be encountered [2]. In our patient, the initial symptoms were an acute headache with confusion. For spinal pachymeningitis, the signs are varied, ranging from simple spinal pain to spinal cord compression [2]. The causes of pachymeningitis are numerous; they can be of infectious, inflammatory or neoplastic origin. Pachymeningitis can also be secondary to idiopathic hypotension syndrome of the CSF (table n ° 1) [2]. It is idiopathic if all etiological investigation are negative. Behcet's disease was considered as a probable etiology of pachymeningitis, but our patient does not meet the classification criteria for Behcet's disease [3]. However, these criteria have not been validated in children and may appear later. Dural biopsy was not performed since the patient remained asymptomatic after a 12 month follow-up, and it is an invasive procedure which is not without risk. Lumbar puncture must always be performed. High protein level in the CSF is almost constant, as was the case for our patient. Boukari et al [4] reported a series of 28 observations of pachymeningitis in adults in which the lumbar puncture, performed in 20 patients, showed high levels of CSF proteins in 17 cases. Lumbar puncture can indicate an increased lymphocyte count. Apart from infectious and neoplastic etiologies, the study of CSF remains of little contribution [5]. Pachymeningitis is well explored by cross sectional imaging. MRI is the gold standard; it allows not only to confirm the diagnosis of pachymeningitis but also to appreciate its importance and its extension [1]. Eventually, MRI can detect a possible complication: hydrocephalus, thrombophlebitis, internal carotid stenosis, lesions of the cerebral parenchyma, panhypopituitarism [2,5]. The dural thickening appears in isosignal or hyposignal in SpT1 and SpT2 weighted sequences, with a marked enhancement by the paramagnetic contrast product. A diffuse hypersignal in SpT2 weighted sequences has also been described [1]. MRI also detects associated intra-parenchymal lesions, which can point towards the etiology of pachymeningitis [5,6]. The treatment of pachymeningitis depends of the underlying etiology. In our country where tuberculosis is still endemic, anti-tuberculosis treatment may have been justified, but this treatment was stopped after 3 months, as the CSF culture for tuberculosis turned out negative.

CONCLUSION :

While the diagnosis of pachymeningitis is evident through brain imaging, the etiological investigation is difficult. The treatment of pachymeningitis depends of the cause (infectious, inflammatory or neoplastic).

Declaration of conflict of Interest :

The authors indicate that they do not have any conflicts of interest. A consent was signed by parents to publish this case .

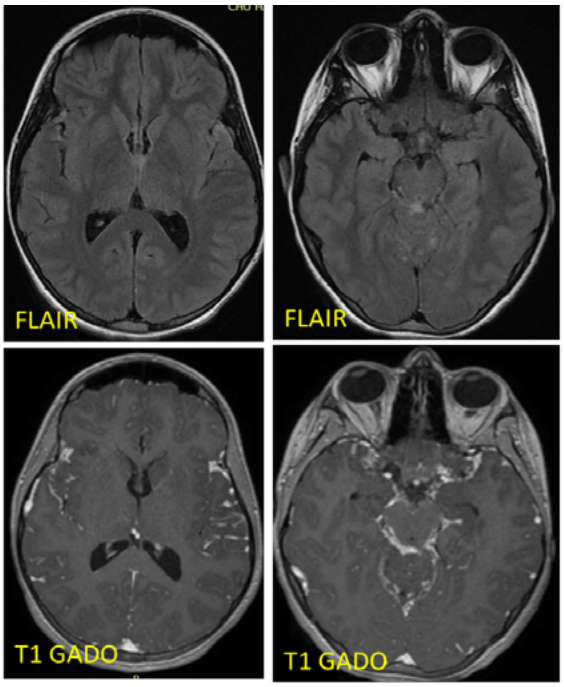

Figure 1 : Cerebral MRI: spontaneous hypersignal on the FLAIR sequence of subarachnoid spaceswith nodular contrast enhancement after Gadolinium injection.

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