Introduction

Chronic subdural hematoma (CSH) is one of the most common disorders observed in neurosurgical practice. The CSH most often occurs in patients \( \geq 60 \) years with brain atrophy, a shrinking or wasting away of brain tissue due to age or disease. In all symptomatic patients with focal neurological deficits surgery is mandatory (1).

The best surgical procedure of CSH is still controversial (2, 3). Most common surgical techniques are a simple one burr hole or, as alternative option, a double burr hole (frontal and parietal). Twist drill craniostomy (TDC) is a less invasive procedure but the craniotomy is often required.

Internal haemorrhagic pachymeningiosis (IHP) is a rare disease characterized by a fibrous thickening and inflammatory infiltration in dural space mimicking chronic subdural hematoma. The pathogenesis of IHP is not entirely clear yet and treatment is still controversial.

Background. Internal haemorrhagic pachymeningiosis (IHP) is a rare disease characterized by a fibrous thickening and inflammatory infiltration in dural space mimicking chronic subdural hematoma. The pathogenesis of IHP is not entirely clear yet and treatment is still controversial.

Patients and methods. The records of five selected cases of IHP histologically confirmed were reviewed, focusing onset, neuroradiology, surgery and outcomes.

Conclusions. IHP is most likely underestimated. Only through multidisciplinary approach it is possible to plane the proper therapeutic strategy. The diagnosis of IHP is confirmed by definitive histology but in some cases is possible with intraoperative frozen section.

KEY WORDS: Internal haemorrhagic pachymeningiosis - Chronic subdural hematoma - Diagnosis - Histology - Surgery.
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Results

In all patients post-craniotomy CT (Figures 2, 3, and 5) showed almost complete disappearance of subdural hematoma.

Definitive histological examination of surgical specimens confirmed IHP. The IHP lesion is clearly subdural even though dural leaflets appeared infiltrated by fibrous tissue. Macroscopically the lesion has hard-elastic consistency and is dark red in color with yellowish patches (Figure 6). Histology shows a fibrous thickening of the dura. Fine vascular network is fragile and bleeding is common. Macrophages with hemosiderin mixed with neutrophils or fibroblasts are frequent (Figures 7 and 8). Around the site of bleeding significant mixed inflammatory infiltrate with fibrin and in some cases calcified areas are present. In the early stages we can find a dissociation of the internal fibrous layer of the dura and vessels hyperplasia, with giants, enlarged, thin small vessels. As the process becomes chronic, new bleedings are possible with haematoma reorganization by granulation tissue formation (5).

The follow up of patients at three years from craniotomy showed total remission of the neurological deficits in 3 patients. Two patients died for other disease: one for renal failure 1 month after craniotomy and another one after 3 months for heart disease.

Discussion

Internal haemorrhagic pachymeningiosis (IHP) is an uncommon lesion characterized by fibrous thickening of the dura mater in which spontaneous or traumatic bleeding can occur. In fact, IHP has clinical evidence when complicated by major bleeding. The brain is flattened and deformed by haematoma. Liquor pressure is often lowered (5, 7).

We don’t know the pathogenesis of IHP but the early stage seems to be the so called pachymeningiosis dissecans, i.e. a relaxation/dissociation of the fibers of the inner...
layer of the dura mater, with the formation of tissue gaps. In these gaps easily bleeding small vessels develop (4, 8). As the process becomes chronic we find, often bilaterally, a stratified fibrous thickening of the dura mater with necrotic material and blood.

As reported by Wepler (6), the pathomorphological substrate of IHP is a proliferative alteration of the fine vascular network of the inner layer of the dura. It is possible that this alteration is preceded or enabled by pachymeningiosis disseccans (6, 8).

In presence of dura mater previously altered (with intradural lesions), the head trauma can lead to the formation of a hematoma but also of a bulky which in any case will be clearly intradural (4, 7). In traumatic chronic subdural hematoma the hematoma is in subdural space, and the organization of the haemorrhage occurs
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Within both of the dura mater and of the leptomeninges (7, 9-12).

In differential diagnosis should also be considered the idiopathic hypertrophic pachymeningitis. This disease is a chronic, fibrosing, non-specific inflammatory process that involves the dura mater of the brain, particularly the falk cerebri and the tentorium. Idiopathic hypertrophic pachymeningitis neuro-imaging mimics small acute subdural hematoma (13, 14).

In many case the cause of IHP is unknow (5). Pre-disposing factors of IHP are: head injury; haemorrhagic diathesis; vitamin deficit; alcohol abuse; intrathecal therapy (methylprednisolone or chemotherapy); complicated meningitis; degenerative disease and collagen va-
scar disease or sarcoidosis; inflammatory conditions, such as cerebral atrophy involving meninges. As reported by Galatioto et al. (7), is critical to differentiate chronic traumatic subdural hematoma from IHP, which is a specific disease entity.

Histologic diagnosis is important for proper therapeutic strategy, if necessary with complete craniotomy.

Conclusions

The IHP is a rare, most likely underestimated and insidious specific disease. IHP must be histologically differentiated from chronic subdural hematoma. The proper treatment is controversial and depends on multidisciplinary collaboration.

References