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clinical practice

Internal haemorrhagic pachymeningiosis: specific disease or complication of chronic subdural hematoma? Report of five cases surgically treated and literature review

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SUMMARY: **Internal haemorrhagic pachymeningiosis: specific disease or complication of chronic subdural hematoma? Report of five cases surgically treated and literature review.**

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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.*

Objective. *We want to emphasize the importance of differentiating pachymeningiosis from chronic subdural hematoma as distinct pathological entities.*

Patients and methods. *The records of five selected cases of IHP histologically confirmed were reviewed, focusing onset, neuroimaging, 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 haematoma - Diagnosis - Histology - Surgery.

Introduction

Chronic subdural hematoma (CSH) is one of the most common disorders observed in neurosurgical practice. The CSH most often occurs in patients ≥ 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. The prevalent hypothesis is that IHP is preceded by an intradural dissociation at the inner layer of *dura mater* called *pachymeningiosis dissecans* (4-8). Clinical and neuroradiological diagnosis and proper treatment of IHP are not yet defined.

The aim of this study is presenting five selected cases of HIP, discussing onset, diagnostic strategy and surgical treatment.

Patients and methods

Between 1999 and 2011, we observed 5 patients (4 male, 1 female; age range 37-88 years, mean 69.2) (Table 1). All patients were admitted to the Emergency Department. Three patients complained of headache associated with hemiparesis, one patient was in coma status, and the last developed only headache without other symptoms. All patients had history of a head injury and three of hypertension. None of the patients had history of bleeding disorder or alcohol abuse.

In all cases preoperative computed tomography (CT) was performed (Figures 1 and 4).

Surgical procedure

All patients had a preliminary drill hole (two times in three patients for the morphologic characteristics of

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TABLE 1

Case	Sex/ Age (yrs)	Previous head injury	Neurological deficits	Site	Surgery	Outcome (3 year follow up)
1	F/88	No	Coma (GCS* 6/15)	Left hemisphere	Two burr holes and craniotomy	Died for heart disease 4 months after surgery
2	M/32	Yes (1 month before)	Left hemiparesis	Right hemisphere	One burr hole and craniotomy	Complete remission
3	M/81	Yes (3 months before)	Right hemiparesis	Left fronto-parietal region	Two burr holes and craniotomy	Complete remission
4	M/74	Yes (2 months before)	Right hemiparesis	Left fronto-temporal region	One burr hole and craniotomy	Complete remission
5	M/71	Yes (2 months before)	Only headache	Left hemisphere	Two burr holes and craniotomy	Died for renal failure 1 month after surgery

** GCS, Glascow Coma Scale*

subdural hematoma, i.e. the presence of fibrous septa) for the evacuation of CSH and redo surgery (craniotomy) to remove the IHP histologically confirmed by the biopsy performed in all cases during the first intervention (Table 1). The excision of the lesion has never difficult.

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.

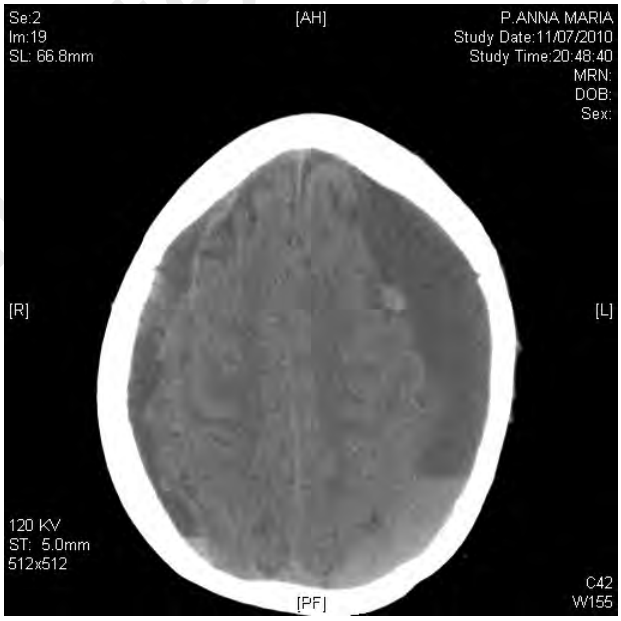

Figure 1 - Case 1. Pre-operative computed tomography.

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

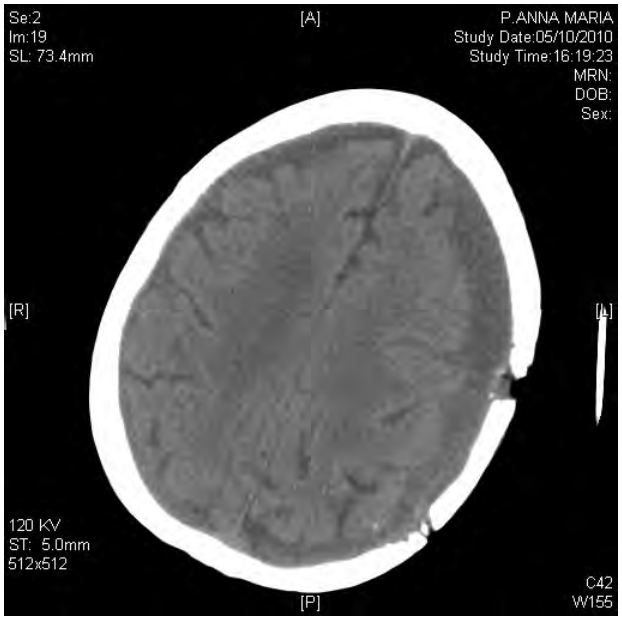

Figure 2

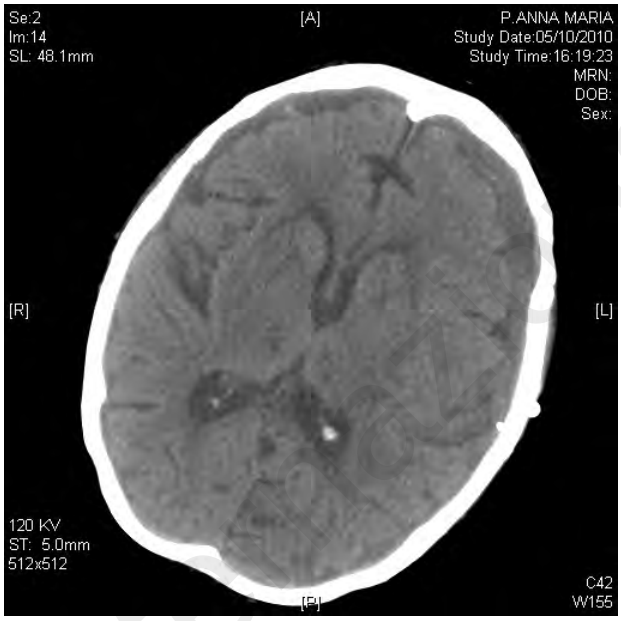

Figure 3

Figures 2 and 3 - Case 1. Post-craniotomy computed tomography shows the disappearance of the lesion.

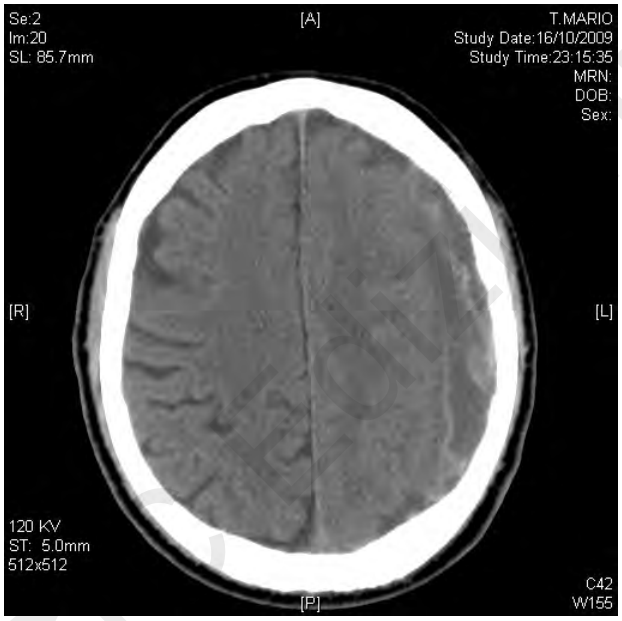

Figure 4 - Case 5. Pre-operative computed tomography shows hemispheric hypodense area in left fronto-parietal region.

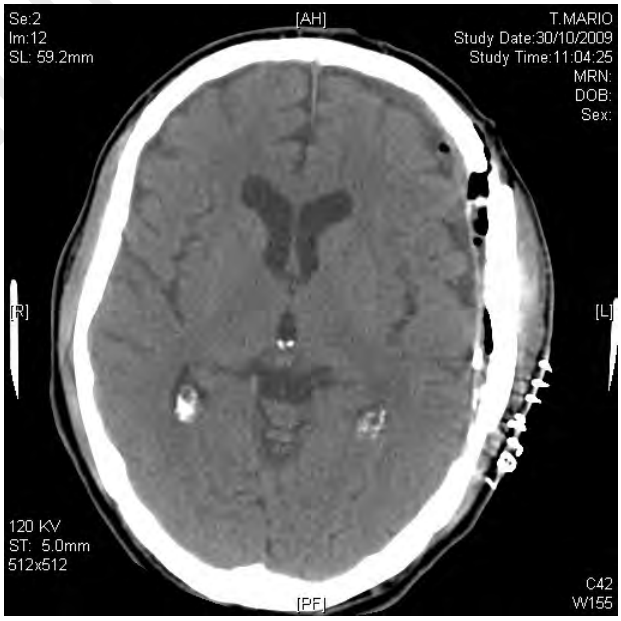

Figure 5 - Case 5. Post-craniotomy computed tomography shows the disappearance of the lesion.

layer of the dura mater, with the formation of tissutal 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 dissecans* (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 haematoma the hematoma is in subdural space, and the organization of the haemorrhage occurs

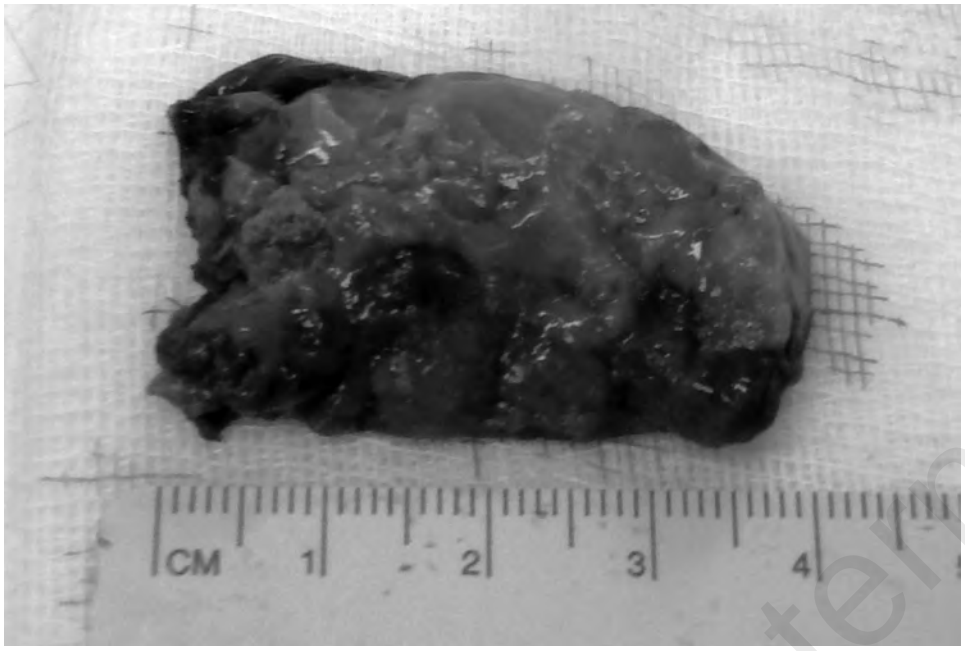

Figure 6 - Case 5. Surgical specimen (40x23mm).

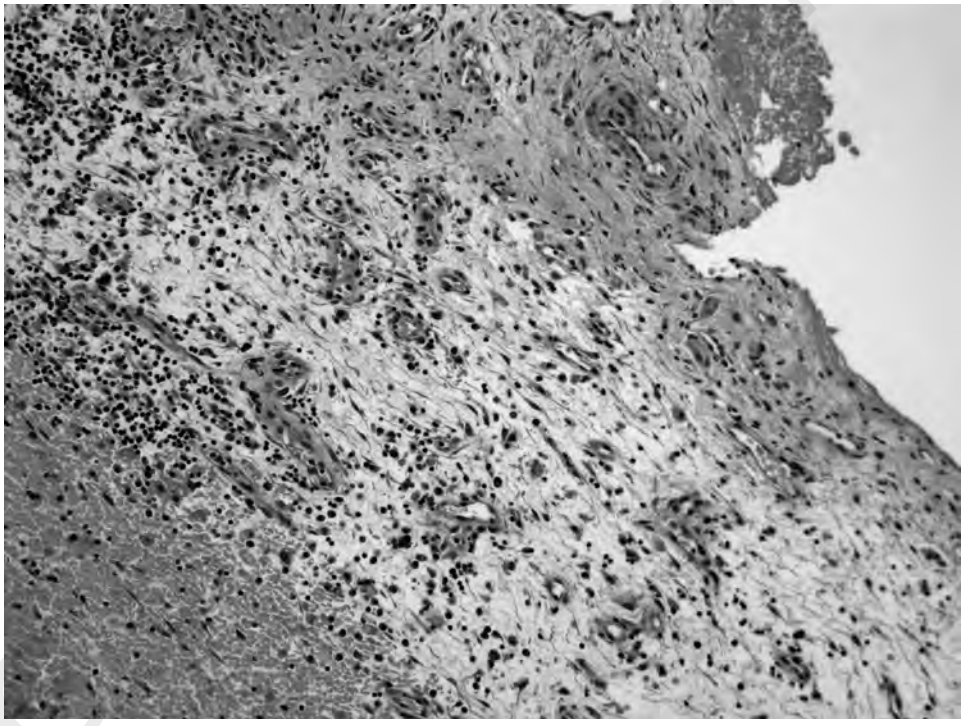

Figure 7 - Histology. Internal haemorrhagic pachymeningiosis. Fibrous thickening of the inner layer of the *dura mater* with thickened small vessels) surrounded by inflammatory infiltrate (H&E, 100x).

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 *falx 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). Predisposing factors of IHP are: head injury; haemorrhagic diathesis; vitamin deficit; alcohol abuse; intrathecal therapy (methylprednisolone or chemotherapy); complicated meningitis; degenerative disease and collagen va-

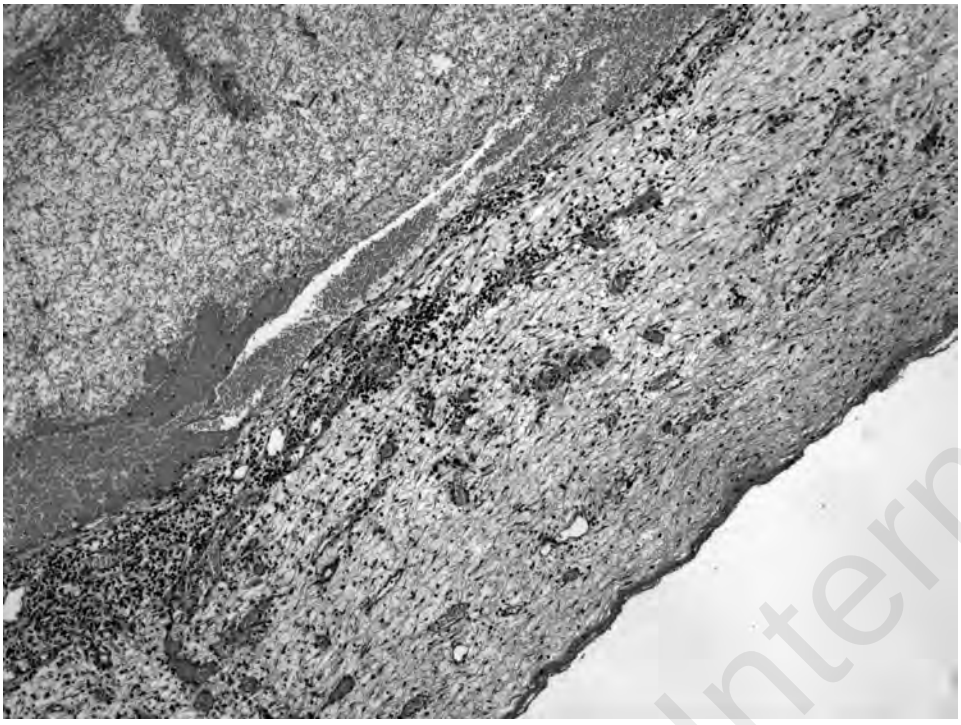

Figure 8 - Histology. Internal haemorrhagic pachymeningiosis. Recent bleeding below the fibrous thickening of *the dura* with chronic inflammatory infiltrate in the meninges (H&E, 50x).

scular 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.

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