Acute necrotic pancreatitis: A rare and not always fatal cause of central pontine myelinolysis

Pancréatite aiguë nécrotique : une cause rare et pas toujours létale de myélinolyse centro-pontine

Introduction

Extra and central pontine myelinolysis (CPM) are considered as osmotic demyelination syndromes (ODS) [1]. The most frequent aetiology of central pontine is rapid correction of hyponatremia. In that case, the prognosis is usually known as highly severe. Nevertheless, a few cases without osmolarity disorder are described [2]. Myelinolysis complicating an acute pancreatitis is extremely rare. To our knowledge, only 2 cases have been described in the literature [3,4]. Mechanisms involved and outcomes of CPM in a context of acute pancreatitis are still unknown. We herein describe, following the CARE guidelines, the case of a patient admitted for central myelinolysis in a context of acute pancreatitis without hyponatremia [5].

Case report

A 31-year old man, with a past medical history of chronic alcoholism, was admitted with severe abdominal pain and vomiting, revealing an acute pancreatitis. Upon admission, neurological examination was unremarkable. Laboratory findings showed an increased serum lipase level at 666 UI/L, sodium level at 133 mmol/L, potassium at 3.50 mmol/L and C reactive protein elevated at 55. Liver enzymes were discreetly increased less than twice normal and bilirubin was normal. Computed tomography (CT) of abdomen and pelvis has confirmed the diagnosis of acute non-haemorrhagic pancreatitis. Considering the medical history of chronic alcoholism with several alcohol rehabilitations, the persistent elevated mean corpuscular volume and the hepatic steatosis seen on CT, the alcoholic origin of this pancreatitis was retained. More, there was no dilation of the bile ducts or cholestasis.

The patient received conservative treatment and was placed on a low-fat diet. In the absence of hemodynamic failure, the patient did not receive any massive intravenous fluid during the initial care that could have corrected a former hyponatremia. One week after admission, the patient rapidly deteriorated with a spastic quadriparesis and disturbance of consciousness. The neurological examination revealed dysarthria, facial diplegia, dysphagia and bilateral Babinski sign. There was no oculomotor abnormality.

The lipase had returned to normal (45 U/L) and the sodium level, assessed extensively with daily control since the admission, had remained within the normal range. A cerebral MRI was performed revealing a hypointense T1 signal without enhancement after contrast, hyperintense fluid attenuated inversion recovery (FLAIR) signal, well limited and symmetric in the central portion of the pontine base (Figure 1). This lesion was associated with discrete lesion in the left striatum. Given the very limited nature of the lesion in the central part of the brain...

**Figure 1**

A. First cranial MRI (T1 and T2 weighted images) realized 15 days after pancreatitis showed abnormal high signal intensity in the central pons. B. A discrete hyperintense signal on the left striatum was also seen.
stem, without oedema or infiltrating character, or without criteria for spatial dissemination, we have ruled out diagnosis of glioma of the brainstem, multiple sclerosis or infection as a listeriosis. Final diagnosis retained was extra and central pontine myelinolysis. He was treated in intensive care unit with conservative care, including thiamine supplement and elective intubation for airway protection during 6 days. The patient remained in critical condition for several weeks, leading to a tracheotomy decision after a week. Repeat MRI at one month showed increased size of the lesions in the pons (Figure 2). The patient’s condition improved spontaneously after 3 weeks with recuperation of a frank mobilization of all members but persistent swallowing disorders. Improvement has continued, leading to decannulation 4 weeks after tracheotomy and discharge 3 months after initial admission in intensive unit care. The patient went back to work 6 months after the event.

Discussion

CPM is a demyelinating process resulting in neurological impairment [1]. If rapid correction of hyponatremia is a classic cause of CPM, acute pancreatitis has been described as a possible cause of CPM [3,4]. The two patients reported had critical condition, leading to death. The scarcity of our case is based on two elements: first the extremely rare aetiology of CPM complicating an acute pancreatitis; secondly, the favourable evolution which is very uncommon in this context.

The genesis of CPM during rapid sodium correction with hypertonic saline implies a hyperosmotic stress that opens the blood-brain barrier (BBB) and consequently creates a vasogenic oedema. This leads to a demyelination of the pontine and extra-pontine glia [4]. Cases of CPM without hyponatremia predominantly occur on a background of chronic medical condition such as chronic alcoholism. Excess production of free radicals is a recognized effect in alcohol use and favours apoptosis [2]. The pathogenesis of CPM after pancreatitis is still elusive. Osmotic injury to endothelial cells by leakage of pancreatic enzymes and increased systemic inflammatory cytokines leading to breakdown of BBB are the main hypothesis. Estrada et al. have suggested that CSF lipase level is directly correlated to the encephalopathy condition [6]. Unfortunately, our patient did not undergo lumbar puncture with lipase assay to support this hypothesis.

The role of alcohol withdrawal, induced by up fasting, has to be discussed. Considering the direct toxic effect and chronic osmotic stress related to the alcohol dependence, CPM should be evoked in any patient with a past medical history of chronic alcoholism presenting with brainstem signs [7,8].

In our case, we are convinced that pancreatitis is the main factor responsible for CPM. Indeed, CPM precipitated by alcohol withdrawal are characterized by a shorter delay of onset of symptoms (2-3 days) with transient progressive course including hallucinations and behavioural changes and less severe condition (no quadriaparesis or comatose state) [2]. Moreover, it tends to occur just after binge drinking associated with dehydration, which is not the case of our patient. Nevertheless, we cannot deny the possible associated pathological effect of alcohol withdrawal, decreased food and water intake in genesis of CPM in our patient. Another diagnosis that can be discussed is pancreatic encephalopathy. This entity refers to a pattern of neurological and psychiatric symptoms complicating acute pancreatitis that occurs on average six days after acute pancreatitis [9]. In these cases, MRI is predominantly normal or depicting aspecific lesions.

According to the literature, chronic alcoholics patients with CPM seem to have a better outcome than cases of rapid correction hyponatremia [2]. An explanation would be that extrapontine part is minor during myelinolysis without osmotic disorders, due to the better basal ganglia’s resistance to osmotic change compared to pontine area. This could explain the favourable outcome of our patient. Strikingly, initial severity was not correlated...
to the good prognosis of our patient. It raises the question of the possibility of inappropriate withdrawal of care in such severe cases in intensive care unit. Although earlier reports have shown high mortality, recent series suggest better outcome with improved recovery. Very few data are available for the most severe patients requiring mechanical ventilation. Recovery is unpredictable on the basis of illness severity. To date, there is no prognostic factor clearly identified. Decisions for the level of care should not be determined by the initial presentation [10,11]. Given that full recovery is possible, intensive care physicians should be aware of this unpredictable outcome and avoid a precipitate limitation of care.

In conclusion, in very rare cases CPM may occur after acute pancreatitis in the absence of hyponatremia and extended conservative care is proposed. In such cases, the evolution may be highly favourable in spite of a severe initial presentation.

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References

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