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Centropontine Myelinolysis Without Hydroelectrolytic Disorders: A Case Report

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Abstract

Centropontin myelinolysis is a mid-protuberant demyelination of various aetiologies. Its genesis is complex, based on several physiopathological hypotheses. The management of this pathology is essentially symptomatic with an uncertain prognosis. We report here the case of a patient with fatal centropontin myelinolysis following acute alcoholism.

Keywords: *centropontin myelinolysis, acute alcoholism, fatal course.*

Introduction

Centropontin and extrapontin myelinolysis (MCP, MEP), also called osmotic demyelination syndrome, correspond to demyelination of the central part of the base of the protuberance, formerly called the bridge, sparing the neurons. This myelinolysis can spread to other structures in the brain.

The first cases were initially reported in the 1950s as a result of chronic alcoholism and malnutrition. It was not until the 1970s that the role of rapid correction of hyponatremia began to be seen.

This clinical entity remains of complex pathophysiology, of poorly codified treatment and is essentially based on general resuscitation measures, with non-specific therapeutic trials and uncertain results. The prognosis is classically poor with high mortality and a significant risk of neurological sequelae.

We report the case of an 18-year-old patient whose only antecedent was a notion of alcoholism following a school failure, who presented with centropontin myelinolysis when his ionic balance was strictly normal.

Observation

We report the observation of Mr. S.A, 18 years old, occasional alcoholic, admitted for afebrile disorder of consciousness.

The examination found a Glasgow score of 10/15 (eye opening at 4, verbal response at 1 and motor response at 5), symmetrical and responsive pupils, without sensitivomotor deficit. Hemodynamically stable (Blood pressure at 120/80 mm Hg and heart rate at 88 beats / min). Respiratory stable, eupneic with 98% pulsed O2 saturation in ambient air. Apyretic at 37.3 ° C and capillary blood glucose was 1.10 g / l.

Faced with this symptomatology, radiological and biological assessments were carried out: A brain scan with a control at H36 was normal, as was the lumbar puncture, thus eliminating the tumor, vascular or infectious origin. The metabolic origin was ruled out before a normal ionogram: (Natremia at 141 mmol / l, calcium at 96 mg / l, uremia at 0.31 g / l, chloremia at 99.8 mEq / l, normal liver function test). Faced with this strictly normal assessment and with the notion of school failure and acute alcoholism, the diagnosis of intoxication was strongly suspected, the toxicological assessment was however not carried out. Patient was kept under surveillance and symptomatic treatment.

The evolution was marked by the improvement of the neurological condition followed by the onset after a six-day free interval of flaccid quadriplegia.

The examination then found a conscious patient, aphasic, symmetrical pupils, with paralysis of the cranial pairs (VII, IX, XI and XII), quadriplegic with lively osteotendinous reflexes. Hemodynamically and respiratory stable one fever at 38 ° C. He was intubated and ventilated due to the swallowing problems he presented.

Brain MRI (T1, T2, Flair with angio sequences) was performed, it had shown a lesion centered on the protuberance **(Figure 1)**, which is increased in size. It is a lesion in T2 hypersignal **(Figure 2)** and nose **(Figure 3)**, without enhancement after injection of contrast product, measuring 24x19x20 mm, without abnormalities in angio sequences and the cervical cord was normal. This aspect was very suggestive of osmotic myelinolysis with pontic localization.

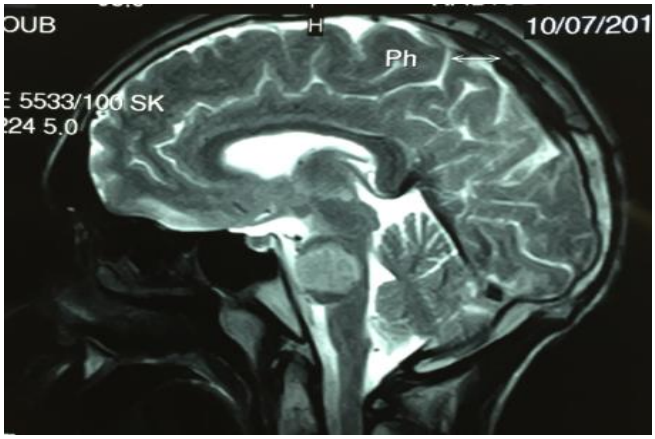

Figure 1: Sagittal T2 view showing a lesion centered on the protuberance.

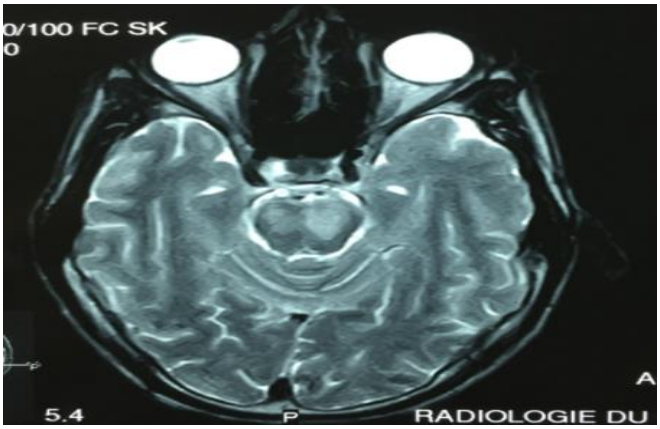

Figure 2: Axial cut with the lesion in T2 hypersignal.

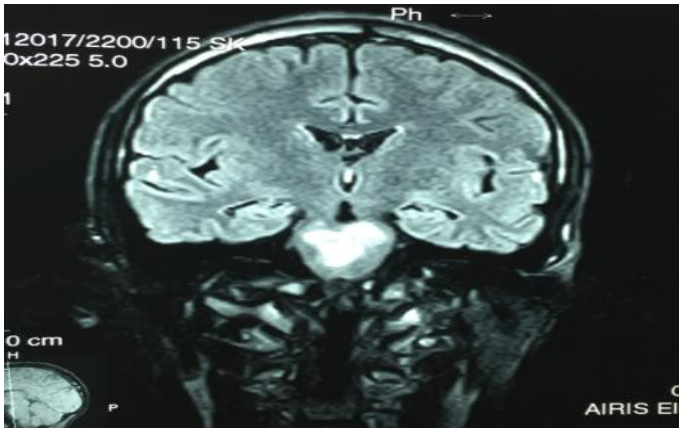

Figure 3: The hypersignal aspect of the lesion in a FLAIR coronal section.

The evolution was marked by the appearance of a plateau fever at 40 ° C, the bacteriological samples came back negative. Broad-spectrum probabilistic antibiotic therapy was started for suspicion of nosocomial pneumonia. The patient died in a table of septic shock. The diagnosis was that of centropontin myelinolysis following acute alcoholism.

Discussion

Originally described by Adams, Victor and Mancall in 1959, centropontin myelinolysis (PCM) is an anatomoclinical entity characterized by the destruction of oligodendrocytes and myelin in the central part of the protuberance, formerly called the bridge, with relative respect for neurons and axons [1]. Other identical histologic lesions mainly affecting the thalamus, putamen, lateral geniculate ganglia and white matter of the cerebellum define extrapontinous myelinolysis (MEP) [2]. These lesions are also called osmotic demyelination syndrome.

The first cases were initially reported to be the result of chronic alcoholism and malnutrition. It was not until 1976 that Tomlinson suspected the role of hyponatremia [3]. Laureno and Karp subsequently confirm the above hypothesis and emphasize the speed of correction of hyponatremia, more than sodium itself [4]. Other studies raise the role of certain pathologies in the predisposition of pontic and extrapontic myelin to osmotic stress, in particular alcoholism, liver disease and nutritional deficits (especially in thiamine, vitamins C and E) during anorexia nervosa. [6,7,10].

Physiopathologically, CPM is most often described in undernourished alcoholics with frequent hyponatremia. The lack of food intake causes maximum reabsorption of sodium and chlorine by the proximal tubule and a decrease in the osmolate flow delivered to the distal level. The excretion of sodium, potassium and urea, on which the excretion of free water depends, is low, causing water retention by residual DHA thus perpetuating hyponatremia [8]. Many complex immunological,

autoimmune and ischemic hypotheses as well as a theory of apoptosis of glial cells have also been proposed, while a purely osmotic mechanism only partially explains the occurrence of MCP [5,8].

In recent years, a new hypothesis based on astrocytic death has emerged after experiments carried out in rats, causing inflammation, micro-glial activation and then demyelination [11]. In our case, the patient was an occasional alcoholic with acute alcoholism following a school failure, but did not present clinical signs of malnutrition or biological stigma of dysnatremia or dyskalaemia [9].

CPM has symmetrical lesions of the center of the protuberance. The clinical course generally occurs in two stages: first, the signs of cerebral distress: agitation, confusion, convulsions, torpor, Cheyne-Stokes dyspnea and possibly even cardiopulmonary arrest. After an initial improvement and a free interval of a few days, neurological signs appear to varying degrees: fluctuation of consciousness, convulsions, hypoventilation, arterial hypotension. In severe forms, pseudo-bulbar paralysis can be observed and be associated with dysphagia, dysarthria, tetraparesis and a picture of locked-in syndrome [8]. Psychiatric forms have also been described [12]. In our case, the patient presented with confusion with an improvement in his state of consciousness corresponding to a free interval of six days after which he presented signs of severity such as swallowing disorders with cranial pair paralysis and quadriplegia.

The diagnosis is suspected in the clinic and confirmed by imaging. Computed tomography is not very sensitive for diagnosis, a hypodense image not enhanced by the contrast product can be observed [13]. MRI remains the examination of choice, showing a symmetrical image of the protuberance in T1 hypointense and hypersignal on the T2 and FLAIR sequences. In our case the initial CT scan was strictly normal, it was the MRI that allowed the diagnosis to be made, showing lesions typical of CPM.

The cerebrospinal fluid study is usually normal, sometimes showing hyperproteinorachia or monoamine metabolites, dosages not performed in current practice [6]. This corresponds perfectly to our case.

The treatment of this rare pathology is poorly codified, based on general resuscitation measures. Therapeutic trials with steroids, intravenous immunoglobulins, Thyrotropin-Releasing Hormone (TRH) or plasma exchanges have been proposed with good results but on small series of cases [8]. Only symptomatic treatment is required, by combating the aggravating factors: correction of hypokalaemia, vitamin substitution, fight against hypoxia, and avoiding rapid correction of hyponatremia. Otherwise, gentle reinduction of hyponatremia has also been proposed [14]. In certain forms of MEP with extrapyramidal manifestations, dopamine could find its place.

The prognosis is variable, with the possibility of recovery ad integrum, or at the cost of moderate or severe neurological sequelae or even death [8]. A large series carried out by Menger in 44 alcoholic patients showed two deaths, recovery ad integrum for one third of the patients, moderate sequelae for one third and heavy sequelae for the remaining third [15]. In another study involving 24 patients, the authors found a favorable prognosis in 60% of cases and an initial mortality of 8% [16].

Conclusion

Centropontin myelinolysis is a serious pathology, most often linked to osmotic aggression following too rapid correction of hyponatremia, or occurring in a particular area of undernutrition or alcoholism.

The diagnosis should be discussed in front of any neurological picture ranging from behavior change to spastic quadriplegia, and should be confirmed by MRI. Even today, there is no specific treatment and the outcome is uncertain.

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