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Case Report

Rare association of central pontine myelinolysis with intrasellar arachnoidocele - casual or correlated?☆

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ABSTRACT

Our case report presents a patient with central pontine myelinolysis and arachnoidocele. He was hospitalized twice these 2 last months for a confusional syndrome associated to an alteration of his general health where metabolic disorders where found: a hyponatremia at 125 mmol/l that was quickly corrected and a hypoglycemia at 0.30 g/l. A central pontine myelinolysis was found as an iso-signal on T1-weighted sequences and a hypersignal on T2-weighted and FLAIR sequences on magnetic resonnance imaging. Central pontine myelinolysis lesions did not enhance with contrast. Incidental imaging findings of arachnoidocele was detected. Through this case, we would like to share with the other practitioners these rare images and the consequence of a diagnostic delay. Indeed, hyponatremia in our patient could be the consequence of the intrasellar arachnoidocele and the overly rapid correction of this chronic hyponatremia caused central pontine myelinolysis, or it is an accidental phenomenon where we found both lesions.

Introduction

Central pontine myelinolysis (CPM) is a part of a group of disorders called osmotic demyelination syndrome, in which damage is caused to different parts of the brain, specially the white matter pontine tracts is the first to be affected [1]. Arachnoidocele is a rare disorder in which the subarachnoid space herniates into the sella turcica, associated with some degree of flattening of the pituitary gland. Our case report presents a patient with CPM and arachnoidocele obvious on the magnetic resonnance imaging (MRI). Though this case, we would like to share these rare images with the other practitioners and discuss if they are a casual discovery or correlated.

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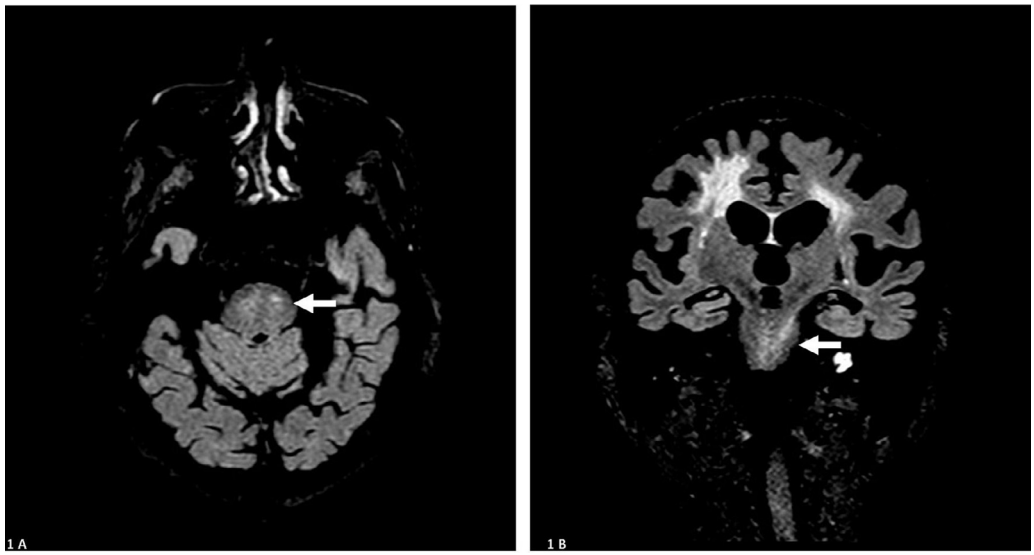

Fig. 1 – Axial (A) and coronal (B) sections of brain MRI In fluid-attenuated inversion recovery (FLAIR) sequence: Hyper signal abnormalities at the center of the pons.

Case report

This is the case of a 78-year-old patient who is known to have a non–insulin-dependent diabetes associated to a chronic end-stage renal failure on hemodialysis.

On questioning, we knew that 6 months ago, the patient had reported nausea, vomits, asthenia, and recurrent syncopes. Hyponatremia had also been found during this period, but no exploration of the etiologic cause has been performed.

He was also hospitalized twice these 2 last months for a confusional syndrome associated to an alteration of his general health where metabolic disorders where found: hypoglycemia at 0.30 g/l and hyponatremia at 125 mmol/l that was quickly corrected. He presented an ischemic stroke a month ago.

In front of these elements and the persistance of the confusional syndrome, an MRI was performed. A central pontine myelinolysis was found. The centropontine lesions appeared as an iso-signal on T1-weighted sequences and a hypersignal on T2-weighted and FLAIR sequences (Fig. 1). Central pontine myelinolysis lesions did not enhance with contrast (Fig. 2). Incidental imaging findings of arachnoidocele was detected on MRI (Fig. 3). We also observed triventricular hydrocephalus, leukoaraiosis, cortical, and subcortical atrophy on the MRI.

Discussion

CPM is a neurological disorder known to be caused by the damage of the myelin sheath of brain cells resulting in demyelination of the pons [2]. It is a rare neurologic condition most frequently caused by the rapid correction of chronic hyponatremia (the development of hyponatremia during more than 48 hours) [1]. Chronic hyponatremia is the result of conditions that cause nutritional deficiencies and electrolyte imbalances such as alcoholism, cirrhosis, renal failure, cancer, syndrome of inappropriate ADH secretion, and liver transplants [1,3].

Some rare associations of central pontine myelinolysis with other disorders have been reported such as Marchiafave-Bignami disease [4] and infantile tremor syndrome [5]. In our case we have a combination of CPM and intrasellar arachnoidocele in the same patient. As far as we know this is the first reported case of such an association. This could be a fortuitous finding, but we attempted to study a possible link between these 2 entities.

Empty sella is classified as primary (PES) when the etiology is unknown and we have excluded a history of previous pituitary pathological conditions, such as radiation therapy or surgery [6]. PES is a relatively common incidental finding in autopsy and radiological imaging (8%-35% in the general population) [6]. History and physical exam are typically normal in patients with empty sella syndrome knowing that the endocrine function is normally intact [2], though endocrine abnormalities are documented in around 19% of patients. When PES is associated with symptoms, it is called primary empty sella syndrome [6].

Moreover, hypopituitarism due to an empty sella is considered as a rare disorder, whose etiology is usually difficult to prove [7]. Normally, it has an insidiously development, with unspecific symptoms, which often delays the diagnosis [7].

Furthermore, hyponatremia as the presenting manifestation of empty sella syndrome is rare. Our hypothesis is that hyponatremia was the presenting manifestation of empty sella syndrome and the overly rapid correction of chronic hyponatremia which caused central pontine myelinolysis. Another possibility is that empty sella and CPM in our case are not linked, and this is more like to be a coincidence.

An exploration of the pituitary was not done. The patient was lost from sight and explorations could not be performed.

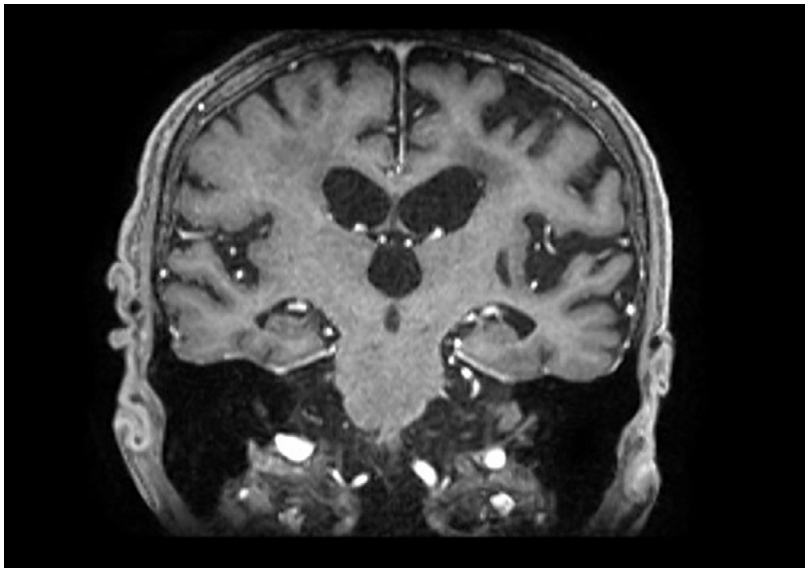

Fig. 2 – Brain MRI resonance with contrast shows no abnormality at the center of the pons.

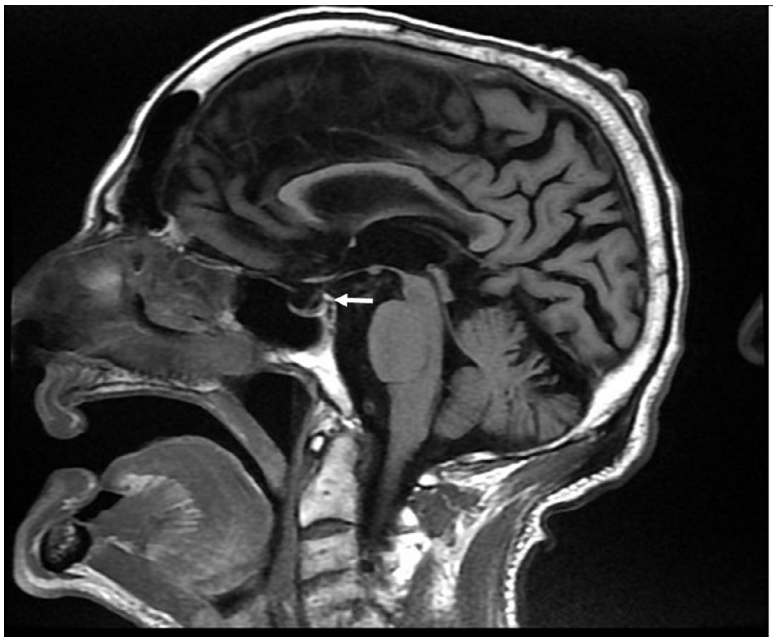

Fig. 3 – Sagittal section of brain MRI in T1-weighted sequence showing an empty sella (intrasellar arachnoidocele).

To the present day, there is no specific treatment for myelinolysis. Preliminary data from animal studies have suggested that reducing the serum sodium in the initial hours and days after rapid correction may be beneficial [8]. Treatment is supportive and aims to prevent complications such as deep vein thrombosis and aspiration pneumonia [9].

Through this case, empty sella and CPM have been reported in the same images which have not been reported in the literature until this day. The main purpose is to avoid diagnostic delay or therapeutic issues.

Conclusion

In conclusion, the best treatment for CPM is the prevention and the early diagnosis. It is important to underline that sodium concentration should be well monitored and fluids and electrolytes titrated carefully. The main purpose if to reduce the chance of involuntary overcorrection in patients with hyponatremia. This case illustrates images of a patient presenting CPM associated to intrasellar arachnoidocele. Hyponatremia in our patient could be the consequence of the in-

trasellar arachnoidocele and the overly rapid correction of this chronic hyponatremia caused CPM, or it is an accidental phenomenon where we found both lesions. Practitioners should be aware of the possibility of having these both lesions and know how to manage it.

Patient consent statement

Consent for publication has been obtained from the patient

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