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OSMOTIC MYELINOLYSIS ASSOCIATED WITH PSYCHOGENIC POLYDIPSY: CASE REPORT

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Abstract: Goal: To report a very rare case of extrapontine osmotic myelinolysis with a predominantly subcortical pattern. Case description: Patient with psychogenic polydipsia evolving with hyponatremia, correction of serum sodium levels and consequent episode of alteration in the level of consciousness. Years later, magnetic resonance imaging showed lesions characteristic of extrapontine OM. Conclusion: As the clinical presentation of OM is nonspecific and the radiological aspect can be characteristic, knowing the different imaging patterns is of paramount importance to narrow the differential diagnosis for the different professionals who may encounter this type of disease.
Keywords: Extrapontine myelinolysis. Psychogenic Polydipsia. Magnetic Resonance Imaging.

INTRODUCTION

Osmotic myelinolysis (OM) is a rare demyelinating neurological disorder related to sudden osmotic changes. Almost always due to abrupt changes in plasma sodium concentrations, being mainly related to the rapid correction of the patient's hyponatremic state (MARTIN, 2004). Additional factors such as hypoxemia are contributors to the worsening and impairment of the disease.

The main radiological aspect of OM seen on magnetic resonance imaging in general is lesions with signal hyperintensity on T 2 -weighted (T 2 WI) and FLAIR sequences and signal hypointensity on T1, located as follows: exclusively in the central portion of the pons (osmotic myelinolysis bridge: MOP); exclusively in regions outside the pons such as the midbrain, thalamus, and/or basal ganglia bilaterally and often relatively symmetrically (extrapontine osmotic myelinolysis: MOEP); or even a combination of pontine and extrapontine components (MARINHO et al,.2014; NETO et al,. 2007; BRITO et al,.2006). An even rarer form of presentation of MOEP is through cerebral cortical-subcortical involvement, which has already been described in rare studies, either through autopsies (OKEDA et al,. 1986; GOCHT et al,.1987),or magnetic resonance imaging by case reports (TATEWAKI et al,.2012; CALAKOS et al,.2000). The patterns of cortico-subcortical lesions associated with MO, despite being quite rare, are relatively specific, especially in the sub-type of subcortical presentation (TATEWAKI et al,.2012).

As in other brain disorders in which, for example, a biopsy is not feasible, imaging tests, especially magnetic resonance imaging, are fundamental for the diagnostic confirmation of OM cases due to the location, distribution and shape of the lesions as well as the signal in the different sequences.

We report a very rare case of extrapontine osmotic myelinolysis with a predominantly subcortical pattern.

CASE DESCRIPTION

Male patient, 28 years old, from the city of Itaú/RN, with a history of significant intellectual disability and autism since childhood, sometimes with attacks of aggression.

At age 25, psychogenic polydipsia began, sometimes drinking about 20 liters of water per day and, little by little, he began to have episodes of nonspecific fainting spells. After one of these episodes of fainting after massive fluid intake, he was admitted to the regional hospital in the city of Pau-dos-Ferros / RN, having been diagnosed with hyponatremia. He was then treated, evolving with a significant lowering of the level of consciousness for five days, with a presumptive diagnosis of encephalopathy related to the aforementioned osmotic disorder. It evolves gradually

with improvement of symptoms, although convulsive episodes are also reported. At the time, he was unable to perform an imaging exam.

Only three years later, a magnetic resonance imaging of the skull was requested to try to better clarify the case. This MRI showed characteristic findings of extrapontine OM with an exclusively cortical-subcortical aspect, retrospectively confirming the presumed hypothesis suggested years ago. Lesions are predominantly temporal and subcortical and are characterized by hypersignal on T2 sequence (Figure 1 a, b and c) and hyposignal on T1 sequence (Figure 2 a and b). In the FLAIR sequence, peripherally there is hypersignal and centrally there is hyposignal (Figure 2 d and d), compatible with chronic alterations. Magnetic resonance imaging of the pituitary without alterations, excluding local alterations related to polydipsia. Today using topiramate and olanzapine,

DISCUSSION

In our case, hyponatremia was the result of primary polydipsia, that is, a psychological disorder that resulted in high water intake by the patient. And correction of hyponatremia presumably would have led to OM. In addition to psychogenic polydipsia, other causes of hyponatremia have been described: alcoholism, malnutrition, adrenocortical insufficiency, SIADH, dehydration due to vomiting, diarrhea or diuretic therapy (MARTIN, 2004; NETO et al,. 2007; TAKEI et al,.1987).

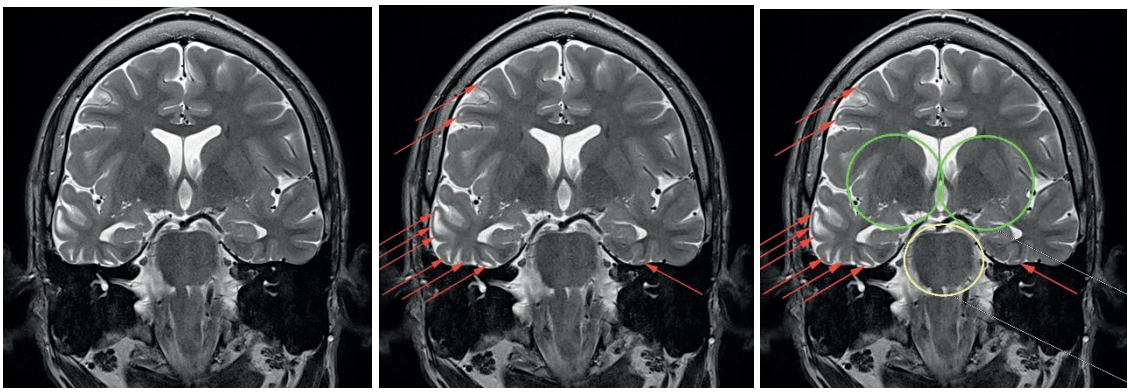
According to Okeda et al (1986), osmotic myelinolysis can be divided into 3 groups: Pontine MO (MOP) with absent extrapontine lesions; combined form of pontine and extrapontine lesions (MOPE); and the exclusive extrapontine form MOP, that is, without pontine lesions. And as a cause, they specifically present the electrolyte disorder, mainly hyponatremia followed abruptly by hypernatremia (OKEDA et al,.1986). This characteristic was observed experimentally in animal models and through the analysis of clinical records.

Gocht et al (1987) examined the distribution of lesions in 58 autopsy cases with OM and found cerebral cortico-subcortical lesions in 15%. As far as we know, OM-associated cortico-subcortical lesions were not clinically familiar, and their imaging findings were only very rarely reported (TATEWAKI et al,.2012), with no reports of isolated cases of subcortical cortical findings being found.

Okeda et al (1986) evaluated the pathological aspects of cortico-subcortical lesions in cases of autopsy with OM, being able to divide the findings into two distinct subtypes of lesions: cortical and subcortical. The cortical form showed laminar cortical astrocytosis and necrosis. The subcortical form presented demyelinating lesions in the adjacent white matter in deep layers of the cortex and mammillary bodies. These subcortical lesions were associated with large areas of vacuoles in the peripheral regions, as clearly demonstrated in our case report (Figure 2).

Magnetic resonance imaging, in our case, demonstrated a predominance of subcortical involvement that somehow seems to have spared the most superficial layer of the cortical gyri, as also described by Tatewaki, Y. et al.

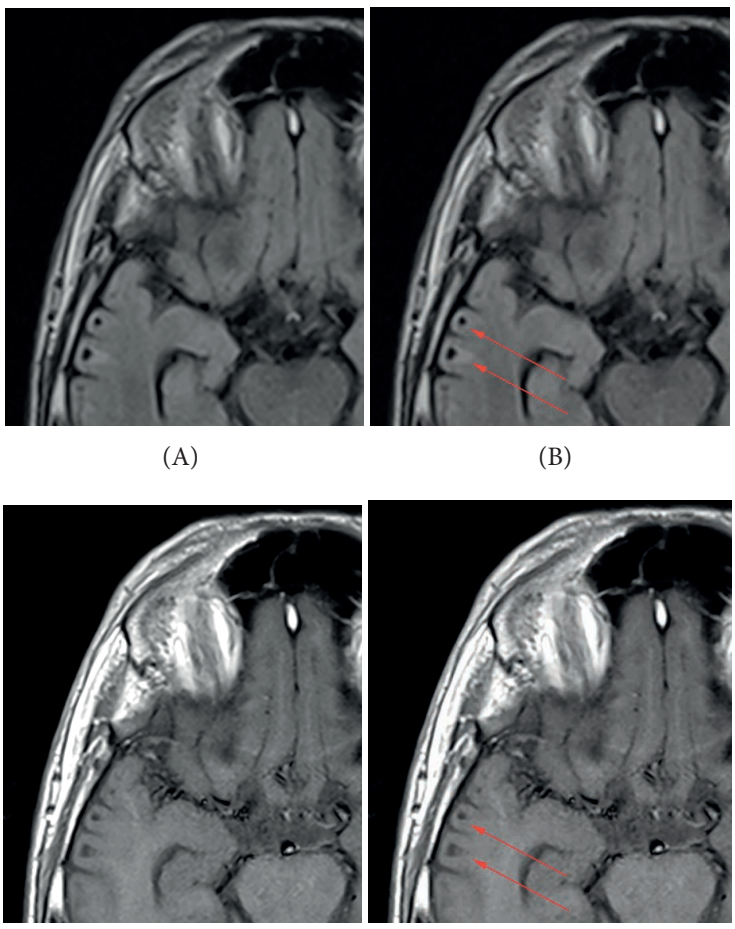
Still regarding the subcortical form, Okeda's group suggested the hypothesis that the proximity and intersection of the white matter with the gray matter at the subcortical cortical junction leads to greater susceptibility to osmotic demyelination as occurs at the pontine base, where there are bundles of white matter intertwined with many pontine nuclei. Thus, they reported that presumably the myelin toxic factors may have been derived from the richly vascularized gray matter,



(A) (B) (C)

Figure 1: Magnetic resonance images of the skull, in the T2 coronal plane, showing predominantly temporal and subcortical lesions ("a", "b" and "c") with arrows pointing to the lesions (in "b" and "c") and the circles (in "c") showing the normal appearance of the bridge (smaller central yellow circle in "c") and the basal nuclei (larger lateral green circles in "c")

*T2 = T2-weighted sequence image.



(A) (B)

(C) (D)

Figure 2: Magnetic resonance images of the skull, in the axial FLAIR plane ("a" and "b") and axial T1 plane ("c" and "d"), respectively without and with arrows pointing to predominantly temporal and subcortical lesions with greater zoom.

*T1 = T1-weighted sequence image. FLAIR = FLAIR-weighted sequence image (fluid-attenuated inversion-recovery).

thereby affecting the adjacent white matter and leading to demyelination. Another factor suggested for the aggravation of the lesions is hypoxia (OKEDA et al,.1986).

The clinical presentations are quite variable and relatively nonspecific, resulting from the different sites where the lesions are affected. Motor changes such as weakness and paralysis can be seen when there is damage to premotor, motor and cerebellar regions. As in other diseases, lesions in the basal ganglia can generate parkinsonian symptoms. Compromise of the pontine region can promote seizures, coma, tetraplegia (MARTIN, 2004). In our case, the presentation was a lowered level of consciousness, in addition to convulsive episodes.

The types of cortical and subcortical involvement have not been clearly distinguished from each other or discussed in the radiological literature with OM. As far as we know, only nine cases with cortico-subcortical lesions have been described in seven articles that include MRI (BOUROUIN et al,.1995; ROH et al,.2009). Retrospectively analyzing these studies, we found that, according to Okeda's pathological classification, the two forms of cortico-subcortical LM lesions can also be discriminated on MRI. Involvement was cortical in four cases (CALAKOS et al,.2000; TAKEI et al,.1987; ROH et al,.2009), subcortical in four cases (TATEWAKI et al,.2012; BOUROUIN et al,.1995; ODIER et al,.2010) and mixed in one case (TAKEI et al,.1987).

In the cortical-type acute phase, curvilinear contrast-enhanced T1 enhancement can be observed (TATEWAKI et al,.2012; CALAKOS et al,.2000; TAKEI et al,.1987), in addition to hypersignal on diffusion-weighted sequences. In the acute phase of the subcortical type, the post-contrast enhancement seemed to be less common (BOUROUIN et al,.1995).

The follow-up magnetic resonance of these OM cases after years showed that there was a predominance of cortical atrophy in forms with a greater cortical component, sometimes with similarities to cortical laminar necrosis in hypoxia (TAKAHASI et al,.1993). In the follow-up of the subcortical form, atrophic lesions were observed in this location, as seen in our case.

The distribution and alterations of the lesions in our case were identical to the type of documented subcortical lesion (OKEDA et al,.1986; TATEWAKI et al,.2012). Of the types of cortical and subcortical involvement in OM, the cortical type appears radiologically and pathologically similar to cortical laminar necrosis in hypoxia, including distribution of the laminar lesion in the cortical layers, in addition to macrophage deposition and demyelination (CALAKOS et al,.2000; ROH et al,.2009)

Given this radioanatomopathological similarity of the cortical form with a hypoxic lesion, it is inferred that the form of subcortical lesion could represent an even more specific form of presentation for OM.

CONCLUSIONS

As the clinical presentation of OM is nonspecific and the radiological aspect can be very characteristic, knowing the different image patterns and their subtypes is of paramount importance to narrow the differential diagnosis, thus being able to help the various professionals who may encounter this type of disease. condition, such as neurologists, psychiatrists, emergency physicians, intensivists, among others.

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