IMAGEM EM NEUROLOGIA/IMAGE IN NEUROLOGY

Pontine and Extra Pontine Myelinolysis Secondary to Hyperglycemia

Mielinólise Pôntica e Extrapôntica Secundária a Hiperglicemia

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Case Report

A 23 years-old female, diagnosed with type 1 diabetes, presented to the emergency department with acute-onset gait and upper limb ataxia, and left-sided hemiparesis. She had a history of poor adherence to the anti-diabetic treatment with HbA1c >14% and significant glycemic fluctuations, between 80-500 mg/dL, the week before admission.

On examination, she was dehydrated. Her muscle tone was normal, she had a mild left hemiparesis and left hemiataxia. Her gait was ataxic with a negative Romberg test and no somatosensory abnormalities.

Her plasma glucose level was 370 mg/dL, with normal sodium and potassium levels. The total osmolality was 295 mOsm/dL. She was treated for her hyperglycemia with insulin therapy.

Brain magnetic resonance imaging (MRI) revealed one central pontine lesion and two symmetrical lesions of the middle cerebellar peduncles with increased signal intensity on T2 and FLAIR, with restricted diffusion (**Fig.s 1 and 2**).

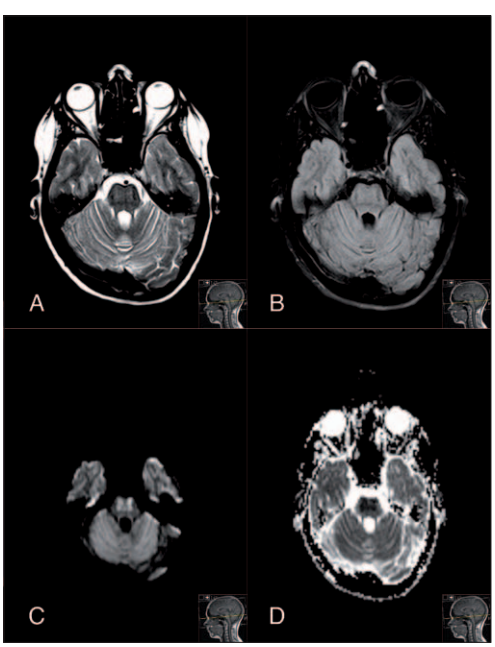

Figure 1. Axial MRI of the central pontine area - Hyperintensity signal in central pontine area in A) Axial fluid-attenuated inversion recovery image; B) T2-weighted image; C) and D) DWI and ADC showing restricted diffusion.

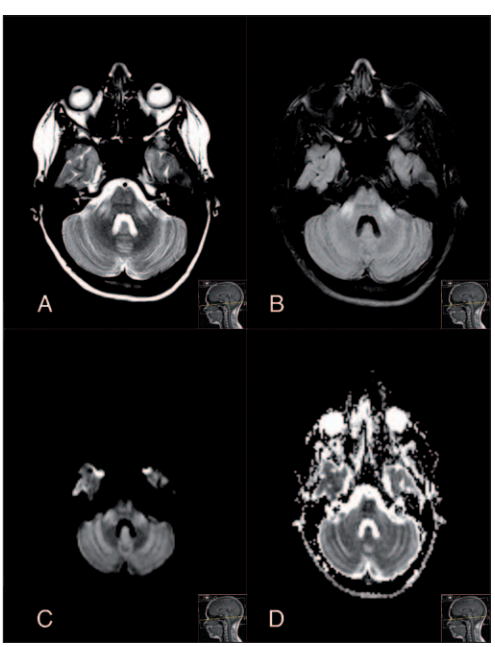

Figure 2. Axial MRI of the middle cerebellar peduncles - Hyperintensity signal in both middle cerebellar peduncles in A) Axial fluid-attenuated inversion recovery image; B) T2-weighted image. C) and D) DWI and ADC showing restricted diffusion.

Her clinical history and neuroimaging were consistent with pontine myelinolysis (also known as osmotic demyelination syndrome) due to hyperglycemia and/or glycemic fluctuations.

She initiated physiotherapy and instructed to maintain strict glycemic control.

Six months after discharge, she had recovered almost completely remaining with mild gait ataxia.

Discussion

Osmotic demyelination syndrome (ODS) is a disorder caused by damage to the myelin sheath of brain cells. This syndrome includes pontine myelinolysis (PM) and extra pontine myelinolysis (EPM), either alone or in combination.[1] The preferred location of osmotic myelinolysis is in the pons, which makes PM much more common than EPM.

EPM is most frequently located in the mesencephalon, thalamus and basal ganglia.[1] The clinical presentation of EPM and PM correlates with the size and the location of the lesions.

PM is characterized by progressive lethargy, tetraparesis, dysarthria, ophthalmoplegia, dysphasia, ataxia, and reflex changes. Clinical symptoms of EPM are more variable.

Diagnosis can be confirmed by finding the demyelinating brain lesions in brain MRI or in computed tomography (CT) images, though with much less sensitivity.[1]

ODS is a severe medical condition, and if left untreated, can lead to a poor prognosis.

Although it is most commonly described after the rapid correction of severe hyponatremia, rarely, it can be found as a complication of hyperosmolar states, like in diabetes mellitus.[2]

PM associated with a lesion of the cerebellar peduncles (secondary to Wallerian degenerescence or due to a primary myelinolysis process) as a complication of hyperosmolar hyperglycemia is rare. ■

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