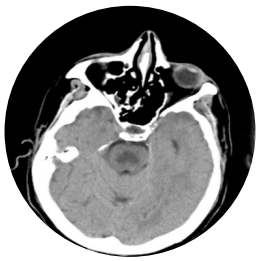

Osmotic demyelination syndrome, a case report

Síndrome de desmielinización osmótica, a propósito de un caso

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Summary

Osmotic demyelination syndrome, previously known as central pontine myelinolysis, is a known disorder in patients with severe hyponatremia in whom rapid sodium correction is performed. It is clinically described as a pseudobulbar palsy, comprised of tetraparesis, encephalopathy, rigidity, ataxia and abnormal movements. It consists of a non-inflammatory demyelination secondary to severe neuronal edema at the pons and other extrapontine locations. It is a very rare pathology, with a poor prognosis and whose only treatment is rehabilitation. A case of a 51-year-old man with fast progressive neurological deficit following rapid correction of severe hyponatremia is presented. The patient required orotracheal intubation due to clinical deteroriation and was diagnosed by computed tomography (CT) and confirmed by magnetic resonance imaging (MRI).

Key words (MeSH)

Myelinolysis, central pontine
Neuroimaging
Magnetic resonance imaging

Resumen

El síndrome de desmielinización osmótica, antes conocido como mielinólisis central pontina, es una enfermedad que ocurre en pacientes con hiponatremia grave en los cuales se realiza una corrección rápida del sodio. Clínicamente, se presenta como una parálisis seudobulbar que consiste en tetraparesia, encefalopatía, rigidez, ataxia y movimientos anormales. El síndrome de desmielinización osmótica es una enfermedad desmielinizante no inflamatoria, secundaria a edema neuronal intenso que se produce en la protuberancia y otras regiones fuera de la protuberancia. Es una patología muy poco frecuente. Sin embargo, tiene muy mal pronóstico, y la rehabilitación es el único tratamiento eficaz. Se presenta el caso de un paciente de 51 años de edad con cuadro de déficit neurológico altamente progresivo después de la corrección rápida de una hiponatremia, con tetraparesia, encefalopatía y rigidez en los días siguientes. El paciente requirió intubación orotraqueal debido al deterioro clínico. Fue diagnosticado mediante una tomografía axial computarizada (TAC) cerebral y se confirmó el diagnóstico mediante resonancia magnética (RM).

Palabras clave (DeCS)

Mielinólisis pontina central
Neuroimagen
Imagen por resonancia magnética

Introduction

Central pontine myelinolysis is a syndrome characterized by demyelination of the white matter due to neuronal edema secondary to sudden changes in osmolarity in the central nervous system (CNS).

Clinically, it presents as a pseudobulbar syndrome with tetraparesis, encephalopathy, rigidity and, sometimes, it can be associated with extrapontine findings such as ataxia and abnormal movements (1).

Initially, it was described in patients with malnutrition and alcoholism, although it is now known to occur more frequently in patients with severe hyponatremia with sodium correction greater than 10-12 mEq in 24 hours (2).

Central pontine myelinolysis requires for its diagnosis analytical and imaging tests, necessarily documenting the hydroelectrolytic disorder and typical imaging features. The findings are mainly detected in magnetic resonance imaging (MRI), although, when the disorder is in advanced stages, it can also be visualized by computed axial tomography (CT) (3).

At present, the only effective treatment is prolonged rehabilitation therapy, so it is essential to prevent this complication with careful management of the patient with hyponatremia to avoid long-term consequences (4) (Figures 1 and 2).

Material and Methods

We present the case of a 51-year-old male patient with no allergies or medical history of interest and no history of malnutrition or alcoholism. The patient was transferred to the Emergency Department by the emergency team due to a generalized tonic-clonic seizure at home, with clinical disorientation and decline of general condition in the last two weeks. On admission, blood laboratory tests showed severe hyponatremia with sodium (Na) less than 80 mEq/L.

Due to the hydroelectrolyte imbalance, the patient was admitted to the intensive care unit with parenteral replacement with hypertonic saline for the next 72 hours. At the end of the hydroelectrolytic correction, blood Na levels were 130 mEq/L.

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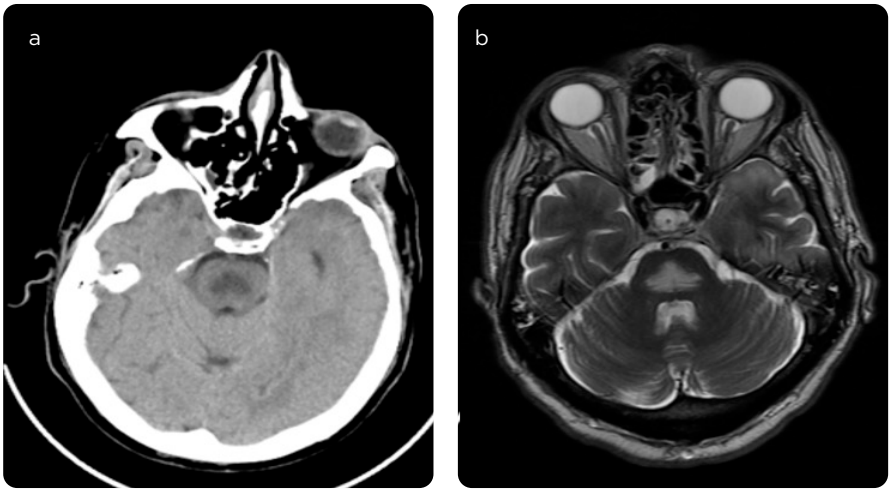

Figure 1. a) CT of the skull without contrast medium showing marked hypodensity in the central region of the pons, with respect to the periphery, and subtle low density in the cerebellar peduncles. b). Brain MRI, axial sequence with T2 information, in a slice similar to CT, showing high signal in the central region of the pons.

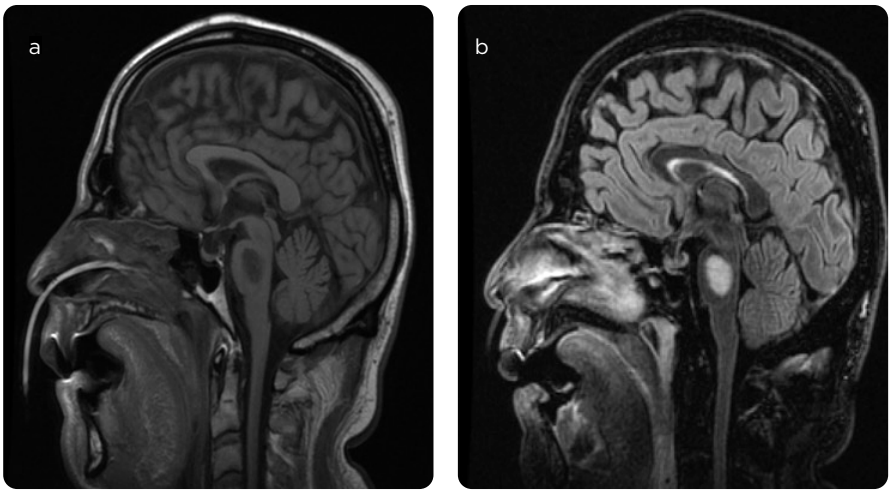

Figure 2. a) Brain MRI, sagittal sequence with T1 information showing marked hypodensity in the central region of the pons with respect to the periphery. b) Sagittal FLAIR sequence with similar findings, confirming an area of high signal in the central region of the pons.

During the days following the hydroelectrolyte replacement, the patient started neurological symptoms consisting of generalized weakness with progression to tetraparesis, encephalopathy with temporospatial disorientation, dysarthria and stiffness in the extremities. The patient deteriorated gradually, his muscle strength and trunk reflexes decreased, requiring orotracheal intubation at 48 hours.

Initially, a CT scan was performed, which showed no lesions, ruling out an ischemic origin of the clinical picture, and the study was complemented with an MRI, which was also normal. Due to the progressive neurological symptoms, treatment was started with dopaminergic drugs, which did not resolve the symptoms. The patient continued with neurological deficit so a CT scan was repeated three days later, with typical findings, which given the clinical context were compatible with a central pontine myelinolysis.

The results were corroborated with MRI, which confirmed the diagnosis of central pontine and extrapontine myelinolysis.

Results

Scan without intravenous contrast medium revealed a very marked low density in the central region of the pons, with respect to the periphery, and a subtle low density in the cerebellar peduncles.

MRI corroborated the findings, with areas of demyelination predominantly in the central region of the pons, as well as in the cerebellar peduncles and basal ganglia bilaterally, confirming the diagnosis of central pontine and extrapontine myelinolysis (3).

Conclusion

Central pontine myelinolysis is a rare but life-threatening clinical entity characterized by a progressive neurological picture that requires a combination of laboratory and imaging data, in which MRI plays a key role (1).

Most cases are associated with accelerated sodium replacement in a patient with severe hyponatremia, although it was initially described in patients with malnutrition and alcoholism (5).

Treatment is currently ineffective; it is based on psychomotor rehabilitation.

References

1. Berry K, Olszewski J. Central pontine myelinolysis, a case report. Neurology. 1963;13:531-7.
2. Laureno R. Central pontine myelinolysis following rapid correction of hyponatremia. Annals Neurol. 1983;13:232-42.

3. Murase T, Sugimura Y, Takefuji S, et al. Mechanisms and therapy of osmotic demyelination. Am J Med. 2006;119:69-73.
4. DeWitt LD, Buonanno FS, Kistler JP, et al. Central pontine myelinolysis: demonstration by nuclear magnetic resonance. Neurology. 1984;34(5):570-6.
5. Perianayagam A, Sterns R, Silver S, et al. DDAVP is effective in preventing and reversing inadvertent overcorrection of hyponatremia. Clin J Am Soc Nephrol. 2008;3:331-6.

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