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Central Pontine and Extrapontine Myelinolysis Complicating a Rapid Correction of Hyponatremia Associated With Hypokalemia in a Pregnant Woman with Hyperemesis Gravidarum with Favorable Outcome: Case Report

Hafida Elmouden*, Mohammed Chraa, Najib Kissani

Neurology department, University Hospital Mohammed VI, Marrakesh, Morocco

***Corresponding author:** Hafida Elmouden
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Abstract **Case Report**

Central pontine and extrapontine myelinolysis is a demyelinating disease of the central nervous system.It is the classical presentation of Osmotic demyelination syndrome (ODS). We report a case of Central pontine and extrapontine myelinolysis in a pregnant patient, who presented electrolyte disturbances due to hyperemesis gravidarum. The patient developed tetraparesis and dysarthria following a rapid correction of hyponatremia.The diagnosis of central pontine and extrapontine myelinolysis was made by brain MRI which showed demyelinating lesions in the pons and in splenium of the corpus callosum. In the reported case, the rapid correction of hyponatremia in it alone does not seem to be the only factor implicated, the rest of the electrolyte disturbances, especially hypokalemia seems likely increased the vulnerability of glial cells to osmotic stress. The treatment was symptomatic. The outcome was favorable marked by regression of dysarthria with partial recovery of motor deficit.
Keywords: Central pontine myelinolysis, Extrapontine myelinolysis, Hyponatremia; Hypokaliemia, hypermesis gravidarum.

INTRODUCTION

Hypermesis gravidarum, or uncontrollable vomiting during pregnancy, is a rare complication of the first trimester of pregnancy that affects various areas of women's health, including electrolyte homeostasis, renal function and may have serious adverse fetal outcomes.

Due to its rich symptomatology and complex pathophysiological mechanisms, hypermesis gravidarum falls within the scope of many medical specialties: gynecology, endocrinology, hepatogastroenterology, psychiatry and neurology [1].

We report a case of centro and extrapontine myelinolysis following a rapid correction of hyponatremia in the presence of hypokalemia in a 35-years-old patient with hyperemesis gravidarum.

CASE REPORT

A 35-year-old woman, 17 weeks pregnant, was admitted to the neurology department for a four-limb heaviness with speech disorders.

Twenty days before her admission, the patient suffered from isolated unstoppable vomiting (without headache, reduction in visual acuity or meningeal stiffness) for which the patient did not consult. Ten days later, the patient worsened. She developed severe asthenia and consciousness disorders with glasgow coma score consciousness disorders at 12/15, which was the reason for his hospitalization at the peripheral hospital before his transfer to the university hospital.

A hydro-electrolytic disorder was incriminated in front of a disturbed biological assessment and a normal cerebral CT: Na +: 124mEq / l, K +: 2.3 mEq / l, Cl-: 89mEq / l, Ca2 + : 84 mEq / l, HCO3-: 41 mEq / l, Total protein: 51g / l. The patient was treated in the emergency department.The control of the biological assessment 24 hours after had shown rapid correction of hyponatremia having passed to Na +: 136mEq / l.The evolution was marked 48 hours later by the occurrence of a motor deficit of four limbs associated with speech disorders reason for which the patient was transferred to our department.

At admission, the clinical examination found a conscious patient, well oriented in time and space, dysarthric with a tetrapyramidal syndrome associating total left hemiplegia, right hemiparesis rated at 3/5, lively deep tendon reflexes in four limbs with bilateral

babinski and pseudo-bulbar syndrome with labioglosso-pharyngeal paralysis and swallowing disorders. Cerebral magnetic resonance imaging (MRI) showed hyposignal with T1 weighting (Fig 1), hypersignal with T2/flair weighting (Fig 2-4) in the protuberance and splenium of the corpus callosum.

Given the obvious context of a rapid correction of hyponatremia secondary to hyperemesis gravidarum as evidenced by the results of the biological assessment with an initial corrected sodium at 124 mEq / L passed in 24 hours to 135 mEq / L and in front of the clinical and radiological data,we retained the diagnosis of central pontine and extrapontine myelinolysis.

Etiological assessment of uncontrollable vomiting showed hyperthyroidism with collapsed TSH: <0.005 µIU / ml (0.15-5), T3: 2.71 µg / ml (2-5.6), T4: 1, 67 ng / dl (0.71 - 1.85); Anti-TPO / anti TG / anti-R-TSH: Negative. Thyroid ultrasound found Thyroid reduced in size hyper vascularized that may be related to thyroiditis.Obstetrical ultrasound showed twin pregnancy at 17 weeks.

Therapeutic management consisted of symptomatic treatment by parenteral nutrition, vitamin therapy, antacid, antiemitic, motor rehabilitation.The evolution was favorable marked by the regression of dysarthria and partial recovery of motor deficit.

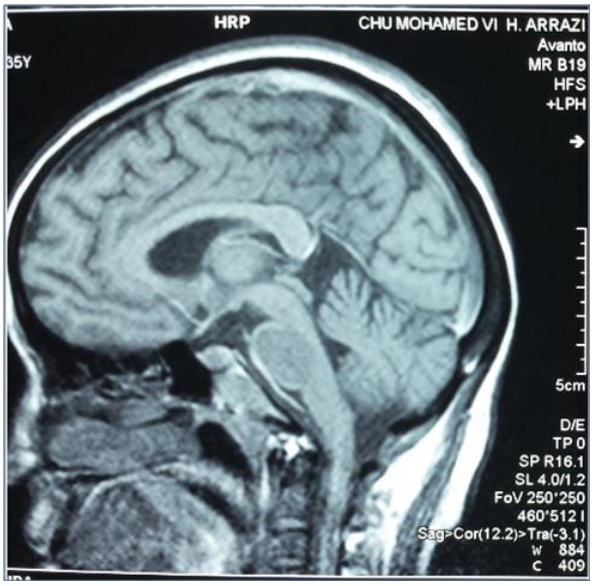

Fig-1: Sagittal T1-weighted brain MRI shows a large hyposignal of the protuberance

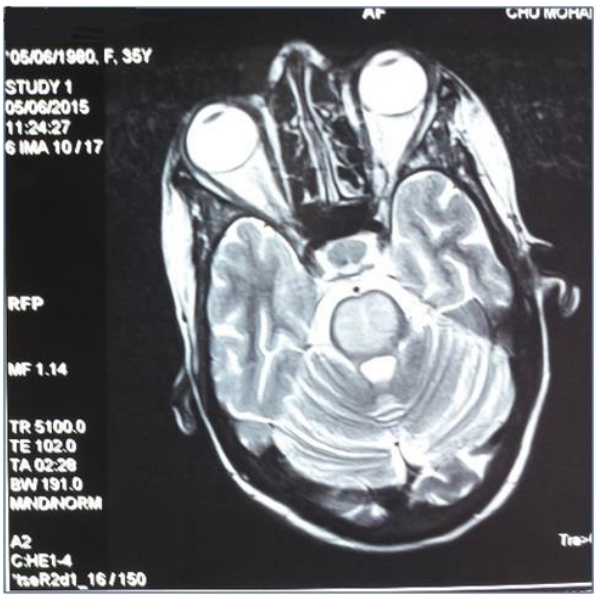

Fig- 2: Axial T2-weighted brain MRI shows a well-defined hypersignal of the protuberance

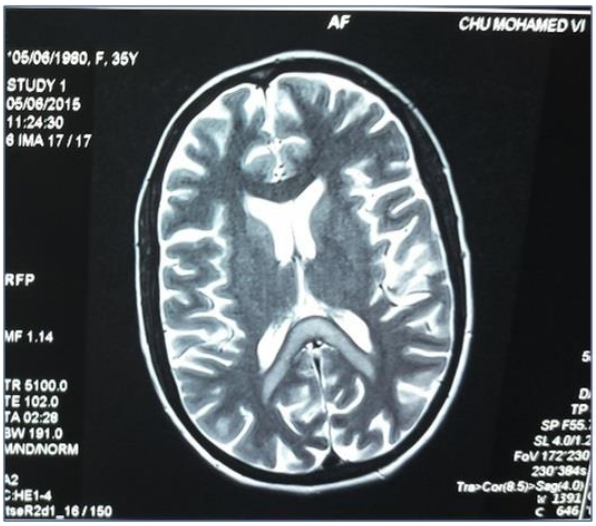

Fig-3: Axial T2-weighted brain MRI shows a hypersignal of the splenium of the corpus callosum

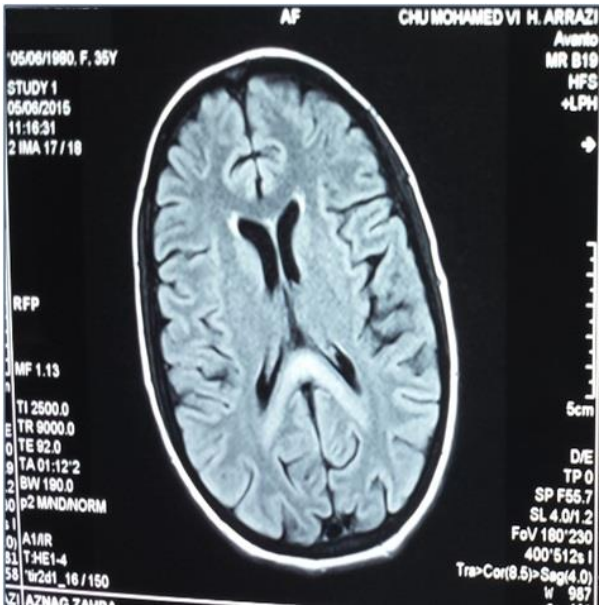

Fig-4: Axial T2-Flair-weighted brain MRI shows a hypersignal of the splenium of the corpus callosum

DISCUSSION

Central pontine and extrapontine myelinolysis corresponds to an anatomoclinical entity characterized by massive destruction of oligodendrocytes and myelin with relative savings of neurons and axons [2]. It is called centropontin when it concerns the central part of the protuberance and extrapontine when it concerns other parts of the brain: the thalamus, the putamen, the globus pallidus, the lateral geniculate ganglia and the white matter of the cerebellum [3].

It occurs in various etiological settings, the most common being the rapid correction of hyponatremia as described in our observation [4-6].Other circumstances of onset of myelolysis have been described in the literature mainly: chronic alcoholism with undernutrition [7-9], deep hypokalemia [10-14].Organ transplantation [15,16], hypophosphoremia [17], lithium toxicity [18], vitamin (C, E) deficiency, extensive burns, hypoxia and malnutrition [19,20]. Recent data suggest that hypokalemia may increase the incidence of central pontine and extrapontine myelinolysis during the correction of hyponatremia, which is the case for our observation [11,13].

The pathophysiology of myelinolysis remains unclear. The most advanced hypothesis is the release of myelinotoxic factors by the vascular endothelium of the cerebral gray matter following the osmotic stress induced by too fast correction of the natremia [21], Nevertheless, the purely osmotic mechanism alone does not explain the onset of central and extrapontine myelinolysis, as it occurs in contexts other than hyponatremia Indeed, Ashrafian *et al*. [22], suggested the theory that myelinolysis lesions in the malnourished alcoholic would be secondary to glial cell apoptosis. The latter is due to a state of chronic energy deficiency causing a dysfunction of Na + / K + ATP pumps

reducing the adaptation of cell volume to changes in plasma tonicity leading to their apoptosis. A new hypothesis has recently been proposed, based on astrocyte death leading to inflammation, microglial activation and then demyelination [23].

Clinically, the evolution of the central pontine myelinolysis is biphasic: the signs of brain suffering are in the foreground with agitation, confusion, convulsions, torpor, dyspnea of Cheyne-Stokes and cardiorespiratory arrest followed 2 to 6 days later by a fluctuation of consciousness, convulsions, hypoventilation, hypotension and in severe forms, pseudobulbar paralysis can be observed and associated with dysphagia, dysarthria, tetraparesis and locked-in syndrome. Extrapontine myelinolysis is classically manifested by abnormal movements: cerebellar ataxia, dystonia, myoclonus and parkinsonian syndrome [19].In the case reported, tetraparesis and dysarthria dominated the clinical presentation and this is explained by the importance of centro-pontine lesions compared with extra-pontine lesions in our patient.

The brain CT scanner is not very sensitive for the diagnosis,it shows symmetrical, centro and extra-pontine hypodensities without contrast enhancement after gadolinium injection [24]. MRI remains the imaging technique of choice. It shows hyposignal images on T1-weighted sequences and hypersignals on T2- and FLAIR-weighted sequences [25].

No curative treatment has been validated to date, therefore, therapies remain symptomatic based on the fight against the aggravating factors recognized by the introduction of a gastric tube or better a feeding jejunostomy, correction of hypokalemia, vitamin substitution and the fight against hypoxia [26].Several treatments have been proposed: treatments with steroids, intravenous immunoglobulins, supplementation with Thyrotropin-releasing hormone (TRH) and plasma exchanges, but no randomized study comparing the various treatments proposed is available at present. There is also no therapeutic consensus [19,27-30]. The treatment then remains preventive, based on a slow correction of the hyponatremias, without exceeding a correction rate of 0.5 mmol / l per hour (10 mmol / l per day) with a multi-day control of the natremia [9].

The prognosis remains reserved and the evolution is endowed with a particularly heavy morbidity [31].A study of 34 patients revealed only 2 deaths, one third having completely recovered another third with moderate sequelae, and the last third with heavy sequelae [32].The peculiarity of our case is its favorable evolution under symptomatic treatment. Lastly, no prognostic factor has been identified until now [19].

CONCLUSION

Central pontine and extrapontine myelinolysis is a serious central nervous system disease with significant morbidity and mortality. The rapid correction of hyponatremia remains the most implicated etiology.In the absence of therapeutic consensus underlines the overriding interest of prevention against this serious disease.

Authors' contributions

All the authors have read and agreed to the final manuscript.

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