Wide Central Pontine, Bulbar and Thalamic Myelinolysis with Sequela

Sekelle Sonuçlanan Yaygın Santral Pons, Bulbus ve Talamik Myelinolisis

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Abstract

Although cases of hyponatremia during pregnancy have been described, it has rarely been possible to show demyelination lesions in central pons, bulbus as well as thalamus, by imaging techniques. We report a case that developed extensive myelinolysis due to the “rapid” correction of hyponatremia as a result of hyperemesis gravidarum. Magnetic resonance imaging showed bilaterally symmetric hyperintense areas in the thalamus and cerebral crus and symmetric hyperintense area in the central pons, with sparing of the rim. In the second day of hospitalization, probably as a result of expansion of myelinolysis, she was intubated and received mechanical ventilator due to bradypnea and a GCS scale of E1M4V2. She was extubated in the 11th day of hospitalization and physical therapy was started. She was discharged in the 30th day of hospitalization with a GCS of 15. But she was tetraparetic (2/5). In the 3rd month of follow up, her quadriplegia improved to paraplegia in lower extremities (3/5). Her follow up examinations in the first year did not change although physical therapies go on. She delivered a normal child.

Key Words: Central pons, Extrapons, Hyperemesis gravidarum, Hyponatremia, Myelinolysis

Introduction

Hyperemesis gravidarum (HG) is a condition of nausea and vomiting that occurs as a result of pregnancy with an incidence of approximately 0.5% of live births. It generally occurs in the first 12 weeks and includes a loss of 5% or more of pre-pregnancy body weight, electrolyte imbalance and dehydration, which occur as a result of excessive nausea, vomiting and the inability to tolerate oral intake [1, 2].

Hyponatremia is the most commonly observed electrolyte disorder in hospitalized patients, but the ‘rapid’ correction of hyponatremia is potentially dangerous. The ‘rapid’ correction of plasma sodium may cause osmotic demyelination syndrome [3]. Although cases of hyponatremia during pregnancy have been described, it has rarely been possible to show demyelination lesions in the central pons, bulbar and thalamus using imaging techniques. We report a case that developed pontine, bulbar and thalamic myelinolysis due to the ‘rapid’ correction of hyponatremia and was discharged with paraplegia of the lower extremities.

Case Report

A 24-year-old woman was admitted to our emergency department (ED) due to nausea, vomiting, fatigue and pins and needles in her extremities. She was 6-weeks pregnant and had no previous history. In her history, she mentioned that she had been admitted to another ED because of morn-
ing sickness four days before where she was diagnosed with hyponatremia (Na: 121 mEq/L) and dehydration. She had been hospitalized for two days and treated with isotonic replacement. In our examination, she was oriented and cooperative. Her Glasgow coma scale was E 4M5V5. There was no meningismus sign, but she had tetraparesis (4/5 for each extremity). Her abnormal vital signs included the following: blood pressure (90/60 mmHg), heart rate (100/min), respiration rate (24/min) and fever (38.8°C). The source of the fever was identified as vaginitis after a foul-smelling vaginal discharge. Her urine analysis showed a pH 6 and ketone ++++. Her biochemical parameters were within the normal ranges (Na: 136 mEq/L, K: 3.49 mEq/L). An arterial blood gas analysis showed the following: pH, 7.47; PCO₂, 29.6 mmHg; PO₂, 59.8 mmHg; SO₂, 92.2%; lactate, 1.2 mmol/L; HCO₃, 21.1 mmol/L; BE, -1.1 mmol/L. Her vitamin B1 and B6 levels were normal. An obstetric ultrasound revealed a viable fetus with normal heart beats in the endometrial cavity, and cranial magnetic resonance imaging (MRI) showed extensive central myelinolysis (Figure 1). Myelinolysis due to the rapid correction of hyponatremia during her previous hospitalization was diagnosed, and she was hospitalized in the neurology ICU. She was administered enoxaparin sodium (Clexane®, 40 mg), methylprednisolone (Prednol®, 40 mg) and a slow saline infusion. Her therapy also included ampicillin/sulbactam for vaginitis. Pseudomonas aeruginosa grew in the culture media, and the antibiotic was not changed. On the second day of hospitalization, most likely as a result of the expansion of myelinolysis, she was intubated and mechanically ventilated due to bradypnea and a GCS scale of E 1M4V2. Her body temperature normalized gradually. She was extubated on the 11th day of hospitalization, and physical therapy was initiated. She was discharged on the 30th day of hospitalization with a GCS of 15. However, she was tetraparetic (2/5). In the 3rd month of follow up, her quadripareis improved to paraparesis in her lower extremities (3/5). Her follow-up examinations in the first year did not change, although physical therapy was continued. She delivered a normal child.

**Discussion**

Hyperemesis gravidarum is diagnosed when prolonged vomiting is present in addition to the inability to tolerate solids or fluids and the presence of ketonuria [4]. Hyponatremia is the most commonly observed electrolyte disorder in patients with HG [3]. A number of risk factors have been identified for the development of central myelinolysis, with the most important being the serum sodium concentration at presentation, also the duration of hyponatremia and the rate of correction is important [5]. No prospective studies have established an absolute safe and definitive speed of correction for hyponatremia. However, conservative therapeutic goals for its correction have been proposed: 8 mmol/L in 24 h, 14 mmol/L in 48 h and 16 mmol/L in 72 h [6]. For our patient, we believe that hyponatremia was corrected too rapidly during her first hospitalization.

The pathologic mechanism of myelinolysis is attributed to the hyperosmotic stress produced by its rapid correction. This induces endothelial injury and damages the blood-brain barrier, resulting in the release of myelinotoxic or oligodendroglial destructive factors [7]. Myelinolysis is most commonly observed in the pontine neurons; however, it may also be observed in extra-pontine sites, such as the thalamus, the basal ganglia, the internal, external and extreme capsules, the claustrum, the amygdala, the cerebellum and the cerebrum. Demyelination is patchy, noninflammatory, frequently symmetric and avoids the axons of nerve cells.

The clinical presentations of central pontine myelinolysis (CPM) are heterogeneous and depend on the regions of the brain involved. The classical symptoms of myelinolysis are spastic quadripareis and pseudobulbar paralysis, which
reflect damage to the corticospinal and corticobulbar paths. The frequently observed symptoms of this disorder include acute para- or quadriplegia, dysphagia, dysarthria, diplopia, loss of consciousness, and other neurological symptoms associated with brainstem damage [7, 8]. Our patient presented with tetraparesis that progressed to pseudobulbar paralysis and inability to breathe.

No specific treatment is known for myelinolysis [9]. Wakui et al. [9] suggested that TRH might improve neurologic damage by increasing blood flow to the brain. They showed clinical improvement but not radiological improvement. We did not use TRH because further studies are needed. Our patient also showed clinical improvement with supportive care; she was tetraparetic (2/5) at admission but improved to paraparesis in her lower extremities (3/5).

Imaging techniques demonstrate abnormalities in the pontomedullary junction in most patients. In T2 imaging series, confluent hyperintensity is observed in the central pons with sparing of the peripheral nerve and corticospinal tracts. The lesion is devoid of a mass effect and contrast enhancement. The effect of the illness on the cerebellar cortex is not typically observed using MRI. The regions most commonly affected are the lentiform nuclei and the thalami in extrapontine myelinolysis. Occasionally, supratentorial white matter structures such as the internal capsule and corpus callosum may be involved. When patients survive, a slow and gradual decrease in the size of their lesions is noted in follow-up studies, but their lesions do not resolve completely [10].

In conclusion, when patients are admitted to an ED with complaints of neurologic symptoms, such as quadriplegia and pseudobulbar paralysis, in the presence or absence of an electrolyte imbalance, clinicians should consider CPM. A detailed history is needed because the electrolyte imbalance in these patients may have been previously treated.

Conflict of interest statement: The authors declare that they have no conflict of interest to the publication of this article.

References
10. Hegde AN, Mohan S, Lath N, Lim CC. Differential diagnosis for bilateral abnormalities of the basal ganglia and thalamus. Radiographics 2011; 31: 5-30. [CrossRef]