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Rare Case of Isolated Extrapontin Myelinolysis

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Abstract

Extrapontine myelinolysis (EPM) is one of the complications occurring secondary to rapid correction of hyponatraemia, and is, along with central pontine myelinolysis encompassed by the more recent term osmotic demyelination syndrome.

We report the case of a patient operated for gastric tumor in whom the post-operative assessment finds a deep hyponatremia quickly corrected causing an apyretic conscience disorder related to osmotic demyelination syndrome confirmed on MRI.

Keywords: MR Features; Extrapontin Myelinolysis; Hyponatremia; Osmotic Demyelination Syndrome

Abbreviations

EPM: Extrapontine Myelinolysis; SIADH: Syndrome of Inappropriate Antidiuretic Hormone ADH Release

Introduction

Extra pontine myelinolysis is a demyelinating process involving areas of brain outside the pons. A rapid correction of hyponatremia the main cause. Extra pontine myelinolysis is lethal, and MRI of brain is the main diagnosis. We report a case 55-year-old patient suffering from sudden apyretic conscience disorder following gastric surgery.

Case Report

We report the case of a 55-year-old patient operated for gastric tumor in whom the post-operative assessment finds a deep hyponatremia quickly corrected causing an apyretic conscience disorder with tremors and myoclonus, a cerebral MRI was relayed first showing a symmetrical and bilateral hypersignal T2 FLAIR at the level of the insular cortex (Figure 1 and 2), basal ganglia, with restriction to diffusion (Figure 3) related to extrapontine myelinolysis (Figure 1 and 2)

Discussion

Extra pontine myelinolysis was described first time in 1987. It's a rare, acute, demyelinating process that involves the areas of the brain outside the pons. Central pontine myelinolysis which involves the pons, also known as the osmotic demyelination syndrome. It commonly occurs in association with extra pontine myelinolysis [1].



Figure 1 and 2: Axial section sequence T2 flair showing symmetric and bilateral hypersignal of the insular cortex, basal ganglia.



Figure 3: Diffusion axial section B1000 showing diffusion restriction.

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A metabolic insult to the brain in the form of over or rapid correction of hyponatremia (a change in serum sodium more than 25 mmol/ litre within 48 hours), is commonly the etiology of extra pontine myelinolysis [2]. Moreover, it is said that some predisposing factors attribute to its severity like alcoholism, malnourishment, sepsis, severe burn, SIADH, chest infection etc. Extra pontine myelinolysis is more commonly found in chronically hyponatraemic patients in whom it occurs 7 - 14 days after acute osmotic shift [3].

Postural limb tremors, myoclonic jerks, parkinsonian presentation, dystonia, catatonia or pyramidal dysfunction, are the main extra pontine myelinolysis manifestation [3].

On MRI, demonstrates a bilateral and often symmetrical hyperintense signal on T2 and diffusion at the level of the gray nuclei (especially the thalamus, but also the lenticular nucleus and the head of the caudate nucleus), but also at the level of the internal and external capsules, the hemispherical, cerebellar and medullary white matter and at the level of the lateral kneeling bodies [4].

Osmotic demyelination is usually irreversible, it could be lethal, and has no proper management. Thus, prevention is the key. The rate of correction of serum sodium should not be more than 1 - 2 mmol/litre/hour during first few hours and not more than 8 - 10 mmol/lit in first 24 hours.

Conclusion

Extra pontine myelinolysis is a rare demyelinating process caused by a rapid correction of hyponatremia. It involved areas of brain outside the pons. MRI of brain remains crucial to asse diagnosis.

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