

Case report

Extrapontine myelinolysis effects in intracranial langerhans cell histiocytosis: Case report



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1. Introduction

Langerhans cell histiocytosis (LCH) is an uncommon disease with an incidence of 0.2–2.0 cases per 100,000 children under 15 years of age [1]. The frequency in adults is not known. The hypothalamic-pituitary manifestations of LCH (often diabetes insipidus) are well known. Thus, complications with sodium level shifts may be present. Rapid changes of sodium concentrations are associated with osmotic demyelination syndromes including pontine and extrapontine demyelination. Here we present a case of a woman with intracranial LCH and subsequent diabetes insipidus who developed extrapontine myelinolysis after she missed doses of desmopressin and subsequent rapid correction of hyponatremia.

2. Methods

The patient's electronic medical record was reviewed including sodium trends, radiology images, and hospital course.

3. Case description

This is a 66 year old female with history of Langerhans' cell histiocytosis with biopsy-confirmed suprasellar metastases and associated pan-hypopituitarism, who was transferred to our institution with concern for hydrocephalus. She had completed five rounds of radiation therapy and two cycles of chemotherapy, most recently one week prior.

She presented to a community hospital with unresponsiveness. Her sodium was 174, and had been 137 three days prior. There was concern that the patient may have missed desmopressin at her nursing facility and had not been awake enough to drink water to thirst. Sodium was corrected from 174 to 139 over four days with a drop from 174 to 157 within the first ten hours. Her mental status improved to the point where she was awake and following commands. She was transferred to our institution with concern for hydrocephalus as etiology for her presenting unresponsiveness. Presenting sodium was 139. Shortly after transfer she became unresponsive again with exam limited to cranial

nerve function only. She was evaluated with an MRI brain and EEG monitoring; however both were unrevealing for a cause. She had an external ventricular drain placed with concern for hydrocephalus being the etiology for her unresponsiveness without improvement in her exam. She remained comatose and one week later a repeat MRI brain showed patchy areas of diffusion restriction involving the frontal and occipital cortex as well as the external capsules bilaterally with corresponding FLAIR hyperintensities, concerning for extrapontine myelinolysis (Figs. 1 and 2).

When the patient returned to the hospital four months later with pneumonia, she had a tracheostomy and gastrostomy tube in place. Her mental status had improved to the point where she intermittently was awake enough to nod to simple questions otherwise she had minimal spontaneous movement and was bed bound. She was readmitted one month later at which time her neurological exam was unchanged. At last follow up the patient had transitioned to hospice care.

4. Discussion

LCH is a challenging disease and can manifest in many ways ranging from a solitary lesion of bone to a multisystem, life threatening disorder [2]. Central nervous system (CNS) involvement is common and has been reported in 16% of patients [3]. The hypothalamic-pituitary axis is the most frequently involved intracranial region in LCH. The characteristic features on imaging consist of enlargement of the pituitary stalk with potential progression to space-occupying tumors extending to the pituitary and hypothalamus [4]. In LCH with associated diabetes insipidus, MRI can reveal loss of the physiologic hyperintense signal of the posterior pituitary on T1-weighted images, correlating with the loss of antidiuretic hormone-containing granules.

The term osmotic demyelination syndrome encompasses central pontine myelinolysis and extrapontine myelinolysis. The majority of these cases occur when initial serum sodium is < 120 meq/L before correction; however sodium may be higher in patients with diabetes insipidus who have developed moderate hyponatremia with desmopressin and then discontinue the medication. In this situation the

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Fig. 1. Post contrast Sagittal T1 MR shows a lobulated enhancing suprasellar mass which was biopsy confirmed to be Langerhans cell histiocytosis.

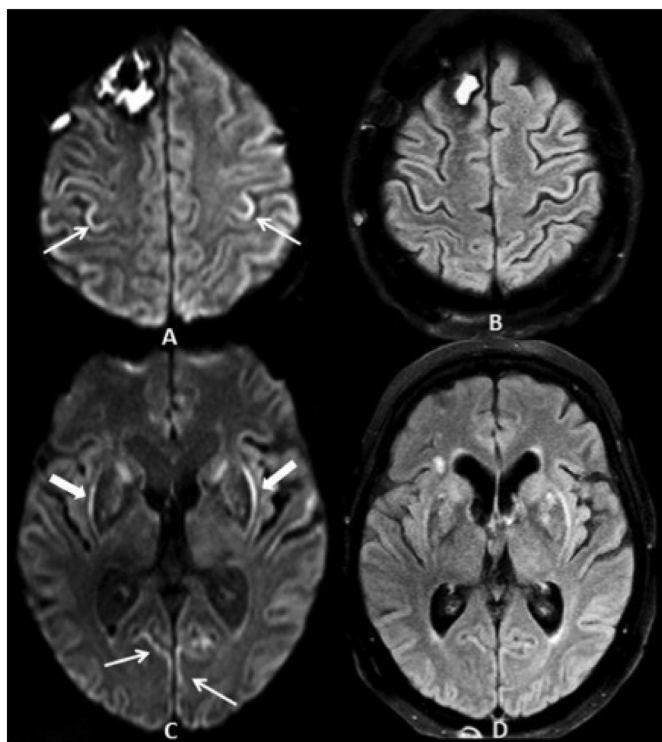


Fig. 2. Axial diffusion images (A&C) demonstrate restricted diffusion in the frontal and occipital cortex (arrows) as well as the external capsules (block arrows). Axial FLAIR images (B&D) show corresponding subtle hyperintensity. The right frontal lobe signal changes are related to prior craniotomy and biopsy.

sodium level may increase rapidly from water diuresis. While osmotic demyelination syndrome is usually associated with rapid correction of hyponatremia; rarely it has been described with correction of hypernatremia.

MRI findings of osmotic demyelination syndrome consist of abnormal hyperintensity involving the pons and extrapontine sites. While extrapontine myelinolysis commonly occurs in association with central pontine myelinolysis; it can also be seen in isolation [5]. The extrapontine sites include the basal ganglia, thalami, and white matter [6].

The clinical course of osmotic demyelination syndrome is typically described as biphasic. Patients may present with severe electrolyte disturbances leading to seizures or encephalopathy. Mental status then tends to improve with correction of sodium; however they can deteriorate 2–3 days later into coma [5]. Osmotic demyelination imaging abnormalities typically lag behind clinical symptoms for up to two weeks [5]. MRI findings in this condition, specifically for central pontine myelinolysis, often consist of a symmetric trident-shaped area in the pons on T2-weighted and FLAIR sequences. Our patient's MRI demonstrated restricted diffusion in the frontal and occipital cortex as well as the external capsules. Axial FLAIR images showed corresponding subtle hyperintensity. The prognosis for osmotic demyelination syndrome is variable; in one study with 34 patients (with central pontine myelinolysis) 2 died, 10 survived with significant neurologic sequelae, 11 had mild deficits, and 11 recovered completely [5].

In summary, our patient had recently been discharged from the hospital on desmopressin for management of diabetes insipidus related to pituitary involvement from LCH; however was unable to take in enough free water on her own leading to significant hypernatremia that developed within three days. She then had her sodium rapidly corrected and subsequently deteriorated clinically. Osmotic demyelination syndrome resulted; however it is unknown if this was due to the initial quick rise of sodium or the subsequent correction of the hypernatremia. This case demonstrates that in patients who remain comatose and have recently had significant changes in serum sodium levels it might be necessary to repeat imaging several days to weeks later to evaluate for the osmotic effects that were not evident on initial imaging.

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