Adult Langerhans' cell histiocytosis as polydipsia - polyuria syndrome in Covid Times

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Background

Langerhans Cell Histiocytosis (LCH) is a rare disease characterized by clonal expansion of myeloid precursors. It presents at any age with various degrees of systemic involvement, and although cure rates are high, long-term complications may affect quality of life. The diagnosis is difficult, and the condition is rarely found in adults.

Case Presentation

A 35-year-old male without prior history. About a year with polydipsia and polyuria. Physical examination shows a small parietal tumor. Blood tests showed normal renal function, electrolyte concentrations, plasma osmolality and the rest of the biochemical tests were normal. Urinalysis showed negative glycosuria and low urine gravity. Due to the Covid-19 pandemic, he was unable to be hospitalized for study. In the ambulatory setting, a modified water deprivation test was performed and stopped in five hours due to hypernatremia. Hormonal measurements showed hypogonadotropic hypogonadism without other alterations. A Brain MRI showed a loss of physiological hyperintense signal in the posterior pituitary gland, pituitary stalk thickening and a right parietal lytic lesion; a chest CT scan showed thin-walled cysts with a slightly lobulated and irregular morphology, measuring up to 7 mm, in the upper lobes. Treatment with DDAVP was initiated, with good response. A biopsy of the parietal lesion showed positive results of immunohistochemical analysis for S-100, CD68, CD1 and langerin. BRAF V600E was negative. The patient started chemotherapy with vinblastine/prednisone.

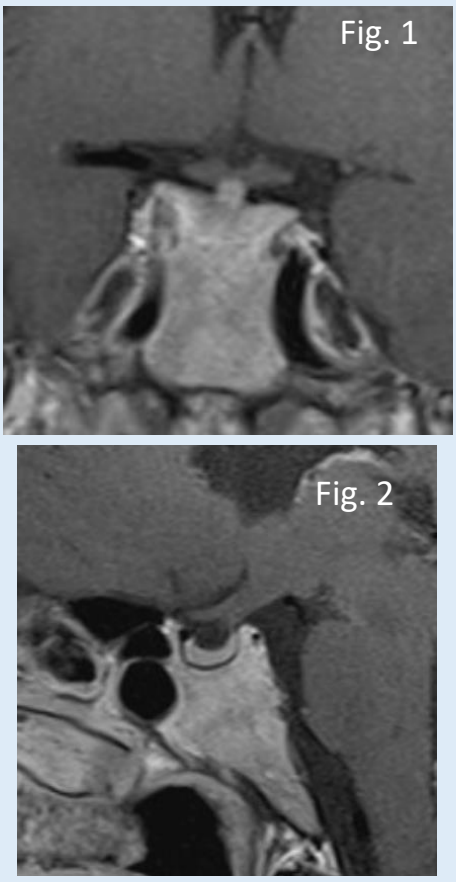
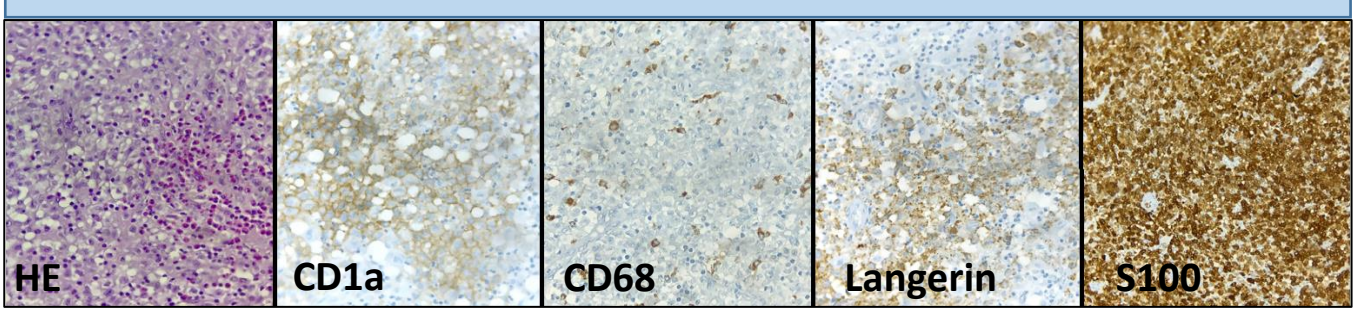


Fig. 1 – 2 Pituitary stalk and clivus involvement

Water Deprivation Test	
Time	Na
09.40	144
11.30	148
13.30	154

Tests	
Cortisol AM	13.2 ug/dL
TSH	3.2 uUi/mL
FT4	0.89 ng/dl
IGF-1	140.5 ng/mL
LH	1.4 mUI/mL
Testosterone	0.496 ng/mL

Inmunohistochemical



Conclusions

LCH has a wide spectrum of clinical manifestations. Given how rare the condition is in adults and its nonspecific presentations, it resulted in a missed and delayed diagnosis. It is classified based on the site of lesions, number of involved sites (single or multisystem/local or multifocal), and whether the disease involves the risk organs. Endocrine manifestations seem to be common and develop mainly in the context of multisystem disease, the most common being central diabetes insipidus. Due to its low prevalence, the prognosis is still uncertain. The current standard of care for treating front-line patients with multifocal LCH or unifocal disease in CNS-risk sites is vinblastine/prednisone.