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【 PICTURES IN CLINICAL MEDICINE 】

Hypothalamic Mass Detected in Langerhans Cell Histiocytosis

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Key words: diabetes insipidus, FDG-PET, Langerhans cell histiocytosis (LCH), hypopituitarism and skin biopsy

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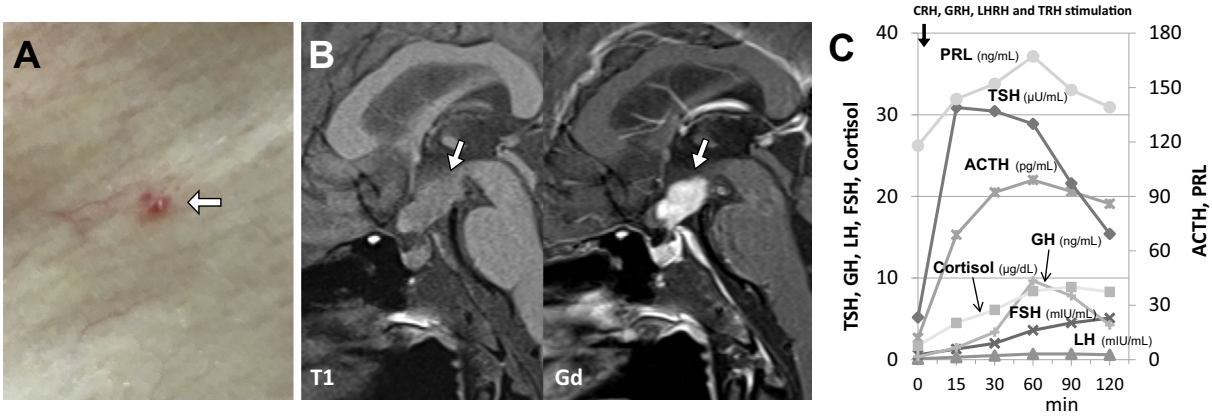

Picture.

A 61-year-old woman suffering from papules (Picture A) was diagnosed with Langerhans cell histiocytosis (LCH) by a skin biopsy. Although the patient was asymptomatic, aside from rashes, fluorodeoxyglucose positron emission tomography (FDG-PET) showed a high uptake in the occipital bone, and endocrine workup demonstrated pituitary dysfunction. Magnetic resonance imaging revealed a tumorous lesion in the basal hypothalamus with gadolinium enhancement and disappearance of the posterior pituitary bright spot (Picture B). Pituitary stimulation tests revealed hypothalamic damage (Picture C). Polyuria and hypernatremia developed after the patient began taking hydrocortisone and levothyroxine, which were improved by desmopressin administration. Hypothalamic damages occur in 10% of central nervous system lesions of LCH (1). The presence of a hypothalamic mass is diagnostic for LCH in patients who had previously been diagnosed with multifocal LCH (2). Central diabetes insipidus with hypopituitarism in LCH is refractory to chemoradiation but requires permanent hormone replacement. Careful screening of systemic lesions, including skin and endocrinological evaluations, are required for patients with asymptomatic LCH.

The authors state that they have no Conflict of Interest (COI).

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