TITLE: Endocrine manifestations in a case of adult-onset Langerhans cell histiocytosis with multisystem involvement.

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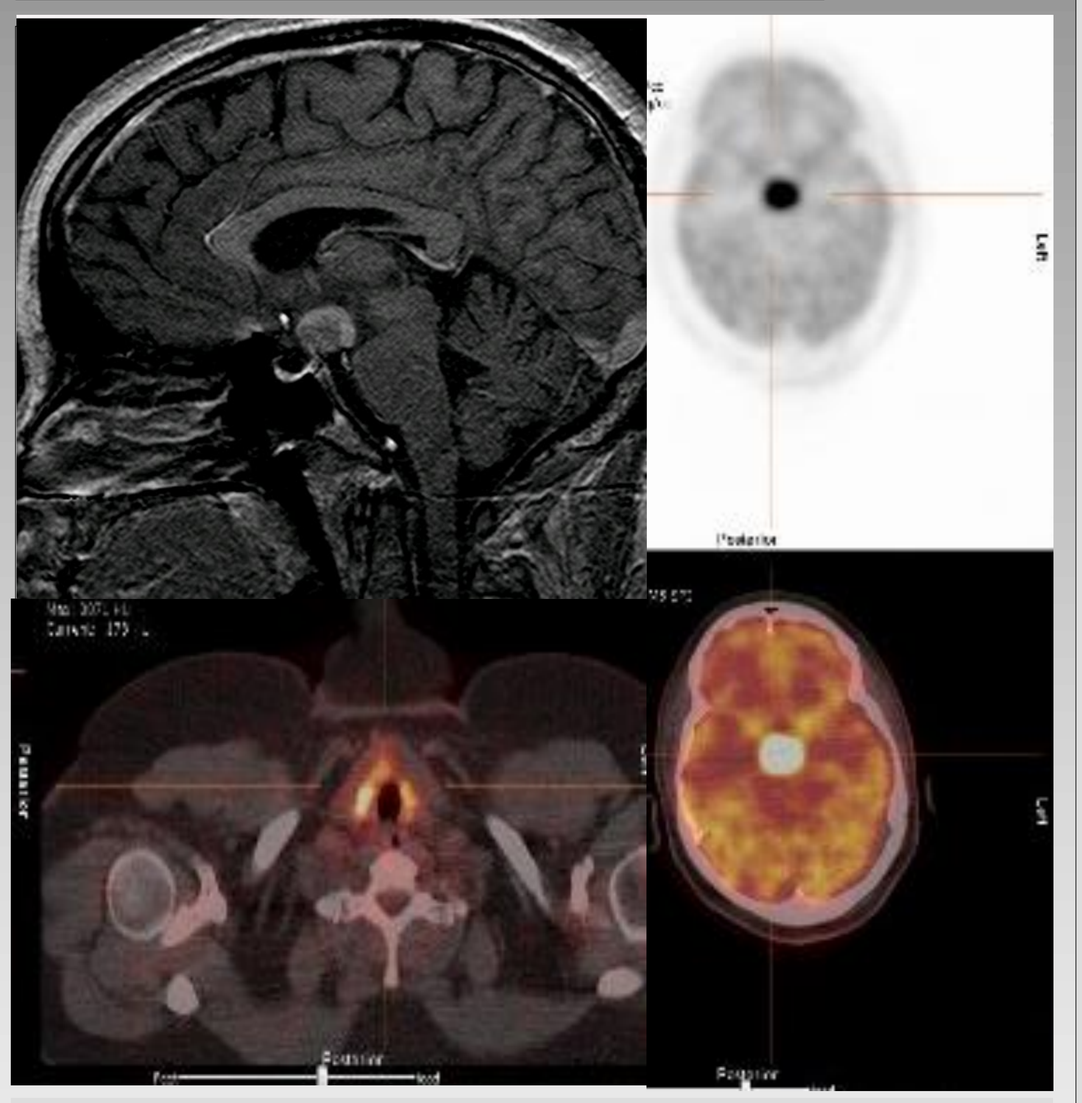
INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disorder with variable clinical presentations. Among the endocrine compromise, the LCH has a predilection for the hypothalamic-pituitary axis, resulting in diabetes insipidus (DI) in up to 50% of cases. However, primary thyroid involvement is extremely rare, and it is usually related to multisystemic disease .



MRI(gadolinium):15x13x22 mm mass involving pituitary stalk and hypothalamic region. 18-FDG PET/CT scan: hypermetabolic pituitary lesion and moderate tracer uptake in the thyroid.

CASE REPORT

We present a 30-year-old woman who requested a medical consultation because of polyuria and was found to have central diabetes insipidus (CDI).

The patient was diagnosed 6 months later with hypopituitarism. Pituitary stalk thickening was detected on magnetic resonance imaging (MRI). Hepatomegaly was also found. Liver biopsy revealed nonalcoholic steatohepatitis with cirrhotic evolution. Three years later she was diagnosed with type 2 diabetes and morbid obesity. Shortly after, she was referred to the hospital because of bradypsychia, somnolence, lost of recent memory, secondary confabulation and dyspnea. Laboratory tests were within the normal range . Magnetic resonance imaging revealed a 15x13x22 mm mass involving pituitary stalk and hypothalamic region. On computed tomography scan, a diffusely enlarged thyroid gland and a 55 mm thymic tumor were observed. Diffuse interstitial lung disease with multiple cysts and hepatosplenomegaly were also detected. Transbronchial biopsy was diagnosed of LCH . Positron emission tomography imaging revealed a hypermetabolic pituitary lesion with a standardized uptake value (SUV) of 24,5 and moderate tracer uptake in the thymo (SUV: 9), thyroid (SUV: 7) and lungs. An electroencephalogram showed mild signs of diffuse brain dysfunction. The results of a bone marrow aspiration were normal and bone marrow biopsy was not succesfully performed due to patient's morbid obesity.

The patient was given chemotherapy with three courses of cladribine and prednisone, showing progressive hepatic and splenic disease and a slight shrinkage of the hypothalamic mass. However, hypopituitarism and CDI were still present. Due to these results cytaribine was added to cladribine. Until now, she has received one course of second line chemotherapy and clinical response has not been achieved.

CONCLUSIONS

In patients with CDI and pituitary stalk thickening, LCH should be considered in the differential diagnosis. Close surveillance for this patient population may be warranted to detect other pituitary hormone deficiencies and even a multisystem disease involving other endocrine glands, such as the thyroid

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