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Pituitary stalk thickening on magnetic resonance imaging as a first sign of Langerhans cell histiocytosis in an adult

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Abstract

Magnetic resonance imaging (MRI) should be performed in the diagnostic workup of central type diabetes insipidus patients to investigate the hypothalamo-pituitary region for a possible thick pituitary stalk. Herein, we presented a 31-year-old male patient who was initially diagnosed as having central diabetes insipidus. MRI revealed isolated pituitary stalk thickening with prominent homogenous contrast enhancement. The remaining findings on cranial MRI were within normal limits for the patient. But a couple of months later he developed some airway symptoms which required a high resolution computed tomography (HRCT) examination. This revealed some suspicious findings in terms of Langerhans' cell histiocytosis (LCH) and following open lung biopsy the patient was diagnosed as having LCH.

Keywords: Diabetes insipidus; Germinoma; Langerhans' cell histiocytosis; Pituitary gland; Pituitary stalk thickening

1. Introduction

Magnetic resonance imaging (MRI) of the hypothalamo-pituitary area is extremely useful in the investigation of patients with central diabetes insipidus (DI). Not only because the hyperintense signal generated by a normal neurohypophysis on T1 weighted images is absent in this condition, but also the precise images that it reveals of the median eminence, pituitary stalk, and anterior pituitary have provided new insights into the etiology of this disease. Indeed, before this technique was used systematically for patients with central DI, no abnormality of the hypothalamo-pituitary area could be detected by computed tomography (CT) scan in a large percentage of patients and most were therefore classified as idiopathic [1].

MRI has revealed isolated pituitary stalk thickening in certain cases of idiopathic or secondary central DI due to infiltrative processes [2,3].

Recently, thickening of the central part of the pituitary stalk, detected by MRI, has been demonstrated [4] in adults with DI and Langerhans' cell histiocytosis (LCH), tuberculosis or sarcoidosis and in children with LCH [5]. Later on, Konrad et al. [6] showed central DI as the first manifestation of neurosarcoidosis in a 10-year-old girl and it has been proven that pituitary stalk thickening can also be caused by neurosarcoidosis in children.

Herein, we describe a 31-year-old male patient who had a central type DI and showed only isolated pituitary stalk thickening on MRI and following a couple of months he developed lung disease which diagnosed as LCH.

2. Case report

The patient was a 31-year-old male who had a history of central type DI and hyperprolactinemia (28.05 ng/dl) for 2 years. Cranial CT examination performed in that time period was completely normal. He was referred to our clinic for MRI examination.

We performed routine brain imaging that was completely within normal limits and after that, we used a pituitary

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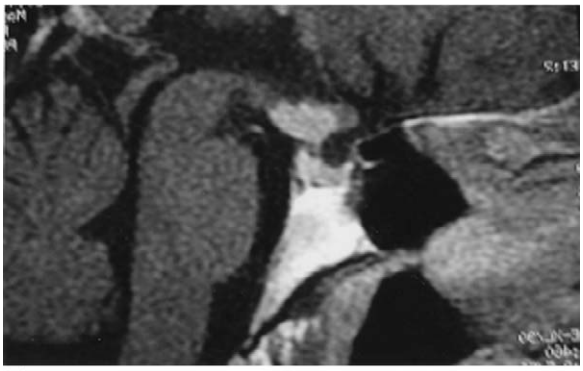

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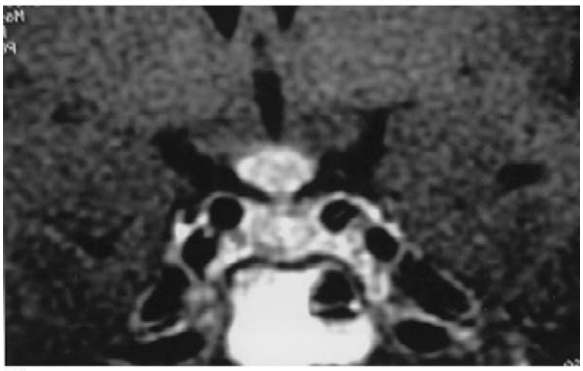

(2)

Figs. 1 and 2. Sagittal and coronal SE T1 weighted postcontrast images, pituitary stalk similarly showing obvious thickening and homogenous contrast enhancement.

protocol in which sagittal and axial SE T1 weighted, coronal SE T1 and FSE T2 weighted and axial, sagittal and coronal postcontrast SE T1 weighted images (following gadolinium administration, 0.1–0.2 mmol/kg) were obtained by using a 1.5 T superconducting magnet. (Picker, Eclipse, USA).

On these images, obvious pituitary stalk thickening was seen and also a homogenous intense contrast enhancement was detected throughout the infundibulum (Figs. 1 and 2). The pituitary gland showed no normal high signal intensity posteriorly. The remaining findings were normal.

The patient was consulted with the neurosurgery clinic to perform a stereotactic biopsy to the infundibulum in order to get histopathological proof but we were recommended to follow up the patient due to the high risk of the procedure regarding the deep anatomic location and size of the lesion.

The patient was followed up by the internal medicine clinic and a couple of months later he had complaints of shortness of breath and fatigue. On physical examination, bilateral crepitan rales were heard along the lower lungs by auscultation. Therefore, a thorax HRCT examination was performed and this revealed interlobular septal thickenings, thin-walled cyst formations and nodular shape thickenings in both major fissures which were characteristic of interstitial airway disease and could be indicating LCH (Fig. 3). The patient was subjected to open lung biopsy and histopathologic examination

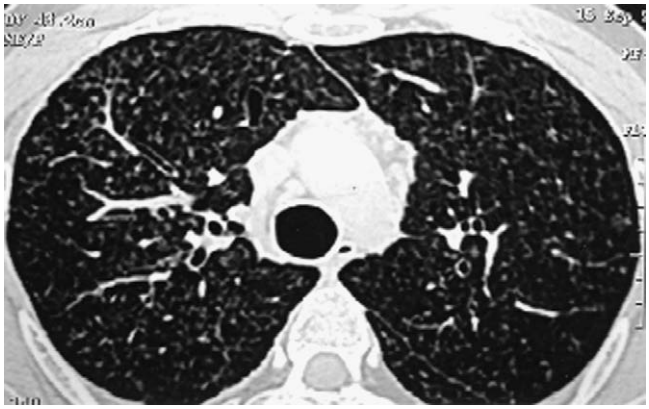

Fig. 3. High-resolution thorax CT axial section, multiple thin-walled cyst formations and interlobular septal thickenings are seen.

confirmed the diagnosis as LCH. The patient was placed on steroids and has also undergone radiotherapy.

3. Discussion

Central DI with isolated pituitary stalk thickening can result from various lesions.

Leger et al. [1] in their study found a precise etiology, which was recognizable at the first presentation in 4 of the 26 patients and these 4 patients had Langerhans' cell histiocytosis (LCH). Also endocrine investigation in these patients showed that anterior pituitary insufficiency of varying severity was present in most of the cases. Although plasma prolactinemia was not evaluated, growth hormone deficiency was the most common abnormality.

In our patient we found hyperprolactinemia.

DI is relatively common in patients with LCH, with a reported prevalence of up to 50% of cases [7,8]. Some of the clinical features of the disease, such as skin and bone defects, are known to spontaneously fluctuate, [9] and in the absence of clinical features, the disease is probably underdiagnosed. Although homogenous enhancement of the normal pituitary stalk with gadolinium is common, these patients demonstrate a pronounced enhancement of the thickened pituitary stalk.

Our patient also had an extensive homogenous pituitary stalk enhancement.

Schmitt et al. [10] in their three children series showed that all the patients presented with acute onset of central diabetes insipidus and isolated pituitary stalk thickening. Within 6 months, two of them developed typical symptoms of LCH manifested with typical peripheric bone lesions.

In our patient who had presented initially with central DI and hyperprolactinemia, pituitary stalk thickening could be regarded as the first typical sign of LCH. As he developed airway disease symptoms a couple of months later and on HRCT some interstitial lung disease findings were detected, this led us to suspect the presence of LCH. Therefore, an open lung biopsy was performed and the patient was diagnosed as having LCH.