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Case Reports

Pituitary apoplexy due to mucormycosis infection in a patient with an ACTH producing pulmonary tumor

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Abstract

Rhinocerebral mucormycosis is an acute, fulminating form of invasive fungal sinusitis occurring principally in individuals who are immunologically or metabolically compromised. The incidence of pituitary apoplexy ranges from 6% to 17%, presenting as a capsule rupture in up to 1.7–2.0%. Isolated cases of mucormycosis are associated with solid tumors and Cushing's syndrome. A 42-year-old, diabetic woman, with Cushing's syndrome of 5 years duration presented with hemiplexy, hemiparesis and altered speech following a syncopal episode and fall. Brain CT scan showed a left temporal lobe infarction. The patient deteriorated rapidly and she died 4 days later. Autopsy findings included: plurihormonal pituitary adenoma with extension to the sphenoid bone and sellar erosion; many thick, septated, mucormycosis hyphae; and recent fronto-temporal brain infarction. Also, a solitary adrenal corticotropic hormone (ACTH)-producing neuroendocrine tumor, 3 cm in diameter, was found in the left lung. This patient illustrates the correlation between ACTH-producing ectopic pulmonary tumor, pituitary apoplexy and mucormycosis.

Keywords: ACTH-producing pulmonary carcinoid tumor; Cerebral mucormycosis; Ectopic Cushing's syndrome; Pituitary apoplexy

1. Introduction

Pituitary apoplexy refers to the clinical picture resulting from hemorrhagic infarction or ischemic necrosis of the anterior lobe of the hypophysis, usually associated with a pituitary adenoma.[1] Pituitary apoplexy is a well-defined clinical entity. Some authors have identified hypoxic pituitary infarction, even in the absence of tumor after delivery; most cases of pituitary apoplexy occur spontaneously, although precipitating factors have been suggested. Magnetic resonance imaging is the investigation of choice.[1]

Rarely pituitary apoplexy is secondary to fungal infection;[2,3] brain abscesses have also been reported. This is uncommon and is also associated with Cushing's syndrome (CS).[4,5] CS results from an increase of adrenal corticotropic hormone (ACTH) due to: excessive adrenal production; adrenal hyperplasia and adrenal tumors either benign or malignant; ectopic ACTH-production due to extra-adrenal tumors (particularly lung, ovary). Extensive radiological and hormonal investigation is necessary to detect extrapituitary sources of ACTH. Differentiation between pituitary and non-pituitary (ectopic) sources of ACTH secretion may be extremely difficult in some patients despite the wide diagnostic armamentarium available.[6]

A 42-year-old, diabetic woman, with CS of 5 years duration, due to an ACTH-producing lung tumor, is presented. Pituitary apoplexy was associated with a plurihormonal pituitary adenoma secondary to mucormycosis.

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2. Case report

We present a 42-year-old woman with a history of hypertension and type 2 diabetes mellitus of 10 years duration, under pharmacological management. Five years before her current illness she developed stress-related lumbar pain and facial and lower limb edema; itching and skin darkening were later observed, and a pituitary adenoma was detected. Surgical treatment was proposed, which she refused. Three years later, she noticed hirsutism, frequent and spontaneous ecchymosis, increase in abdominal girth and weight gain; 2 years before her current illness she noticed progressive decrease of visual acuity.

She then presented with left hemiplexy, left hemiparesis and altered speech, after a syncopal episode and fall.

On physical examination, the patient looked older than she was, with hirsutism, melasma and moon face. She was obese and her blood pressure was 160/100 mmHg. She presented with multiple ecchymosis, violet-spotted skin, dysrhythmic tachycardia (150 bpm), an enlarged abdomen with violaceous striae, asymmetric hypotrophic lower limbs and incoherent speech.

Neurological examination revealed optic atrophy, with facial asymmetry and there was diminished strength and hypoesthesia on the right.

Her hormone profile was thyrotrophin 1.0 microU/mL (0.35–4.94), free thyroxine 9.3 pmol/L (9.0–19.0), thyroxine 50 nmol/L (62.5–150.0), follicle stimulating hormone (FSH) 27.2 mU/mL (26.0–135), luteinizing hormone (LH) 3.4 mU/L (7.7–59.0), prolactin 9.8 ng/mL (3.4–24.1), growth hormone 8.4 ng/mL (0.0–20.0) and morning cortisol 65.5 ng/mL (8.7–22.4).

Four days after her admission, she deteriorated, with anemia and sepsis. Surgical resection of the pituitary adenoma was attempted, but she deteriorated into coma and had a cardiac arrest during surgery.

Fig. 1. Necrotic tissue removed from the sella turcica at autopsy.

2.1. Radiological findings

Brain CT scan showed a cortical hypodense area, which suggested a recent infarction in the left temporal lobe. The sphenoidal sinus had destructive lesions with inflammatory characteristics with wall remodeling that infiltrated the sellar region.

A CT scan of the thorax showed a well-defined, homogeneous, 2-cm nodule in the basal lobe of the right lung, which was also seen on abdominal CT.

An MRA showed narrowing of the superficial branches of the left middle cerebral artery.

2.2. Autopsy findings

The skull base structures showed minor softening, and the meninges were slightly swollen and pale. No abnormalities were found in the coronal sections of the brain. Some friable, soft, whitish necrotic matter destroying the sellar region and the sphenoid sinus was removed (Fig. 1).

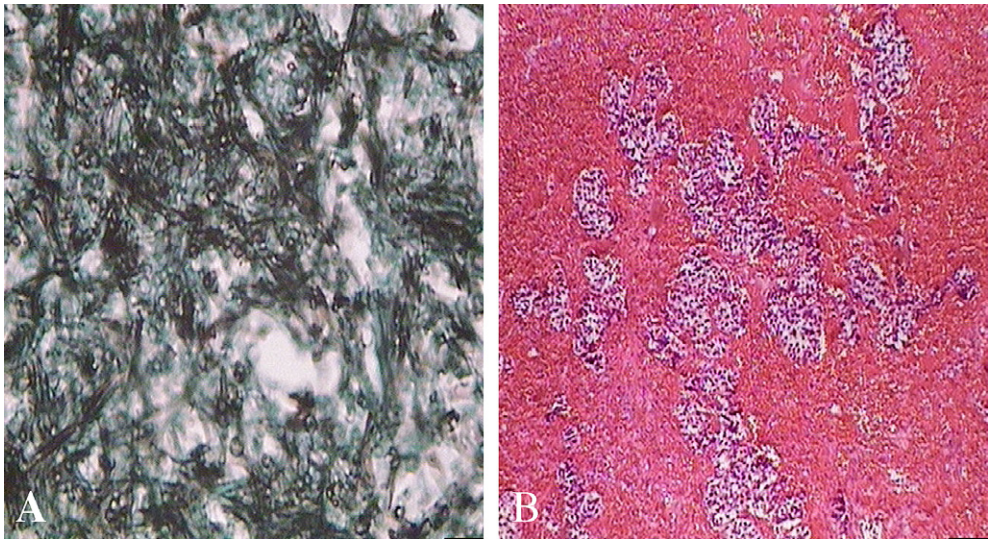

Fig. 2. A: Tissue removed from the sellar region showing necrosis and mucormycosis hyphae (Grocott stain, original magnification × 400). B: Pituitary adenoma with necrotic and hemorrhagic areas consistent with pituitary apoplexy (Hemotoxylin and eosin, original magnification × 400).