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CONTINUING MEDICAL EDUCATION
ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2008, 25(5):691–692

Endocrinology Quiz – Case 4

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A 29 year old woman was referred to the outpatient department claiming that she had difficulty to lose weight. Two and a half years ago, during her pregnancy, she had gained 12 kg. She also reported edema of the face, trunk and the extremities, hirsutism, acne and menstrual irregularities (infrequent and heavy menses), with gradual deterioration during the last year. At this point the patient was advised to loose weight and thyroxine was prescribed, without confirmed hypothyroidism. Because of no improvement of her symptoms and in fact deterioration of the degree of obesity she seeked a second opinion.

On directed questions by the endocrinologist on her second visit, the patient also complained for muscle weakness, easy fatigability, blurred vision, palpitations, emotional lability, irritability, abdominal pain (due to distension), mild sleep disturbances (early morning awakening) and easy bruising. On physical examination the patient was overweight with central obesity (BMI: 29,4 kg/m,^{2} Waist to Hip ratio: 0,95) and suffered from hirsutism (Ferriman-Gallway score: 11). The blood pressure was 140/80 mmHg and the heart rate as 96/min. The patient presented with "moon face", plethora, fill-up of the supraclavicular pits (fig. 1) and "buffalo hump" (fig. 2). Her skin was dry with transparent appearance, generalized cystic acne and lesions compatible with tinea versicolor on the chest and the posterior surface of the neck. The abdomen was distended, painless to palpation with peri-umbilical thin white striae. The patient exhibited prominent difficulty to rise from the seating position, apparently due to weakness of the proximal muscles of the lower extremities.

Given the history of obesity, post partum menstrual

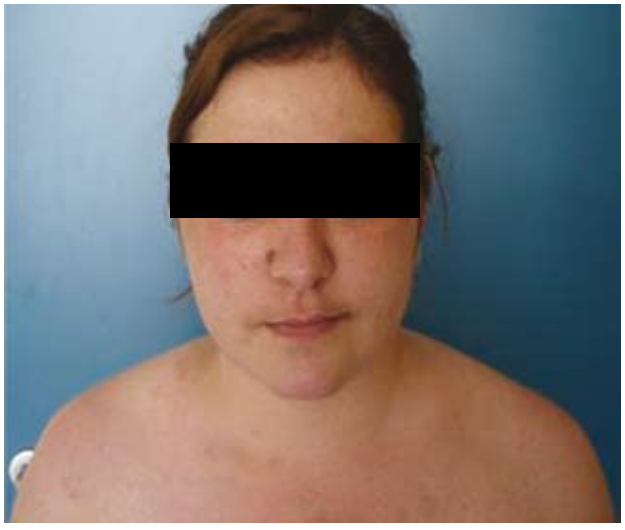

Figure 1

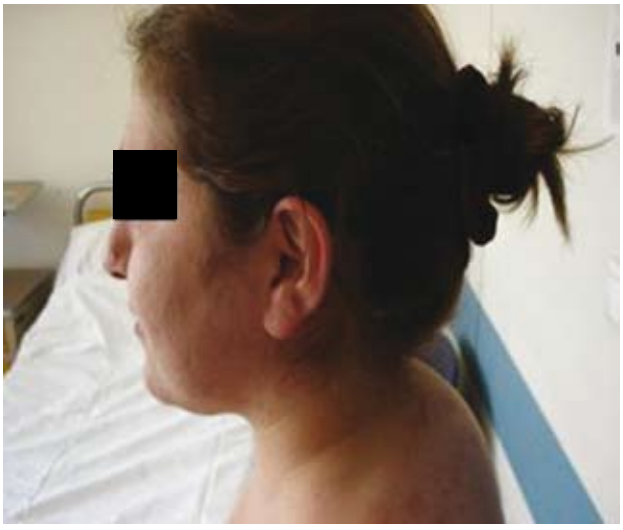

Figure 2

irregularities and edema of the face, trunk and the extremities, which is the running diagnosis?

Comment

Differential diagnosis is depicted on table 1. The laboratory work up of the patient revealed: High urine free cortisol [UFC=1124 nmol/24h (nv: 38–208)], absence of diurnal rhythm of cortisol (8:00= 665 nmol/L, 23:00=693 nmol/L) and undetectable ACTH levels. Additionally, a lack of suppression of plasma cortisol after low (cortisol=529 Nm, nv <50 Nm) and high dose dexamethasone suppression test (baseline UFC=2318 nmol/24h and after suppression UFC=2899 nmol/24h, normal suppression >90% of the baseline). These findings confirmed hypercortisolism and pointed adrenals as the most possible source. Indeed, a CT scan of the upper abdomen was performed which depicted the presence of a mass (d~4 cm) in the left adrenal with smooth margins compatible radiomorphologically with adenoma (fig. 3).

The clinical picture (gradual onset), the laboratory testing (non detectable ACTH) and the imaging study (adrenal mass) confirmed the diagnosis.. Accordingly, the patient underwent a laparoscopic excision of the left adrenal gland and histopathology established the diagnosis of adrenal adenoma. Because the patient´s hypothalamic-pituitary axis and the contralateral adrenal were expected to be suppressed by prolonged endogenous excessive cortisol secretion, she received glucocorticoid therapy both during and following surgery.

Table 1. Causes of Cushing's syndrome

ACTH-dependent Cushing's syndrome	ACTH-independent Cushing's syndrome
- Pituitary adenoma (Cushing's disease) (70%)	- Iatrogenic
- Non pituitary neoplasm (ectopic ACTH) (10%)	- Adrenal neoplasm *Adenoma (10%) Carcinoma (5%)*
	- Primary pigmented nodular adrenocortical disease(<2%)
	- McCune-Albright syndrome (<2%)
	- Factitious

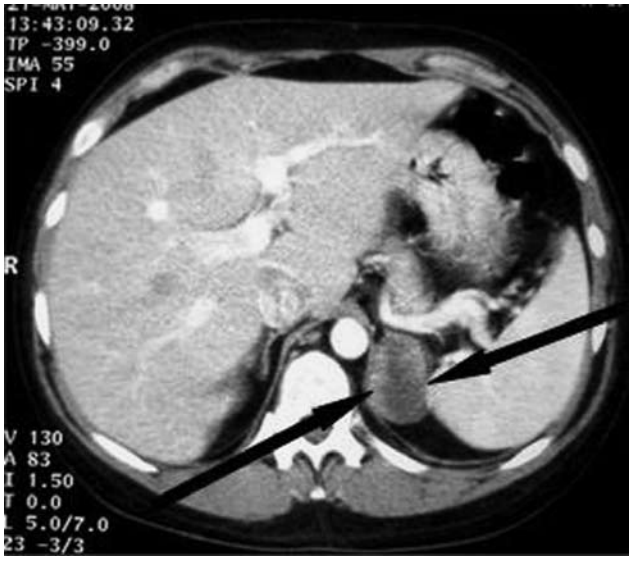

Figure 3

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