

Case Report

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Fatal Factitious Cushing's Syndrome (Münchhausen's Syndrome) in a Patient with a Prolactinoma and Silent Corticotrophinoma: Case Report and Literature Review

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Background

Factitious Cushing's syndrome (CS) is extremely rare condition and its diagnosis is challenging mainly due to a cross reactivity of endogenous and exogenous glucocorticoids in immunoassays. The Münchhausen's syndrome (MS) is a chronic factitious disorder characterized by the intentional production of symptoms to assume the sick role without external incentive [1].

Case Report

A 26-year-old woman was originally evaluated for secondary amenorrhea and galactorrhoea due to macroprolactinoma. Transsphenoidal surgery was performed due to chiasmatic compression, despite the control of serum prolactin (PRL) levels with cabergoline. Immunohistochemistry revealed positive and diffuse staining for PRL and, surprisingly, for ACTH, although she did not exhibit any CS features. After surgery, PRL levels were controlled without cabergoline and no glucocorticoid replacement was necessary. Four years later, the patient presented with typical, albeit sporadic, signs and symptoms of CS paralleled by an intermittent hypercortisolism, indicating potential cyclic CS [2,3]. Exogenous intake of glucocorticoids was strongly denied by the patient. A progressive worsening of clinical features of CS was evident. At that time, laboratory tests (chemiluminescence immunoassay – IMMULITE 2000/SIEMENS, Gwynedd, UK) revealed a markedly elevated urinary total cortisol levels (UTC, up to 12.200 mcg/24 h – nv 50- 310 µg/24 h), elevated midnight salivary cortisol level (0.2- 2.3 µg/dL – nv < 0.12 µg/dL) and normal morning serum cortisol (Fs) concentration. Plasma ACTH was within normal levels (14 pg/ml – nv 10- 46 pg/ml). DHEA-S level was low (208 ng/ml – nv 988 - 3400 ng/ml) reaching suppressive values. Serum PRL levels were normal. Differential diagnosis with pseudo CS by desmopressin test was inconclusive. Abdominal CT showed normal adrenal glands and pituitary MRI exhibited remnant tumor tissue (a small nodular cystic lesion). To further investigate the discordance in UTC and morning Fs, high-pressure liquid chromatography-tandem mass spectrometry (HPLC/MS) was performed and revealed a Urinary Free Cortisol (UFC) level at 6 mcg/24 h (nv 3 - 43 µg/24 h) and a suppressed morning Fs (1.6 mcg/dL – normal 7 up to 25 µg/dL) contrasting with the previous results. The psychiatric evaluation did not identify any mental disorder but the psychological evaluation identified a complex familial relationship, especially about her father, who had alcoholism history and left home since she was a teenager. We identified an ego with a severe fragility, predominating the defense mechanisms of projection, negation, somatization and reactive reactions. When the medical team talked about the procedure of adrenalectomy to control the hypercortisolism, the patient agreed with the treatment even though all the risks exposed, showing a desire to get healthy and to be pregnant as soon as possible. During hospitalization and besides a multidisciplinary treatment approach, the patient showed a persistent hypercortisolism which was associated with high levels of triglycerides, amylase, and lipase. The abdominal CT in this moment showed bilateral

atrophic adrenal glands and a severe pancreatitis with fatal outcome. Additionally, high levels of serum prednisone and prednisolone (7.4 µg/dL and 97 µg/dL, respectively – nv < 0.1 µg/dL) were demonstrated, but unfortunately the results arrived too late. One day before the fatal outcome, her husband finally found pills of prednisone at home.

Discussion

We reported a case of factitious CS with a fatal outcome. The diagnosis of factitious CS represents a challenge to the physician. These patients have high morbidity and increased mortality risk. There are few cases in literature of factitious CS, one of them with fatal outcome. The diagnosis is complex and includes cyclic CS in the differential diagnosis. The difference between these possible diagnoses is the periodicity of hormone fluctuation, which can take years in the case of cyclic CS, but can change dramatically over short periods of time in factitious CS. In our case, variations of 100-to 12.000 µg/24 h in UTC could be observed from one day to the next. In the few cases of factitious CS in the literature, the presence of synthetic glucocorticoids in the blood and urine by HPLC confirmed the diagnosis. Immunoassays for Fs and UTC demonstrated variable cross-reactivity with synthetic corticosteroids and their metabolites. These variations range from 6% for prednisone to 62% for prednisolone, depending on the kit, and can lead to a false suppression of basal cortisol. A reliable ACTH assay is crucial for differentiating ACTH-dependent from ACTH-independent Cushing syndrome. However, the performance of ACTH immunoassays has been questioned in recent studies, in which the results were frequently reported to be within the normal reference interval (i.e., unsuppressed) as in our patient. About the patients' profile in the cases described at literature (17 cases on totally), 16 were women, with a middle age of 39, 9 years, a variable civil state, and 6 of 9 questioned were health professionals or had contact with them. Besides, from 9 patients with a psychiatric evaluation, all of them had some psychiatric diagnosis, including depression, anxiety and bipolar disorder. The classic features of CS were present in 8 cases and some patients already had clinical or surgical treatment for CS [4].

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Conclusion

We described a patient with chronic factitious CS characterizing Münchhausen syndrome with a fatal outcome. Factitious CS is an important diagnosis to consider in the evaluation of patients with apparent hypercortisolism. The main point is the recognition of discordant hormonal data among urinary and serum cortisol, ACTH and DHEAS associated to atrophic adrenal glands on CT scan. In addition, HPLC analysis of blood and urine steroids is the definitive test for the diagnosis of factitious CS and should be performed when there is clinical suspicious of glucocorticoid mishandling.

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