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Clinical case

# Secreting ectopic adrenal adenoma: A rare condition to be aware of

## *Adénome surrénalien sécrétant : une rareté qu'il faut connaître*

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**Abstract**

Ectopic adrenal adenoma causing chronic Cushing's syndrome (CS) is a rare phenomenon. Diagnosis is usually made years after disease onset because of the insidious nature of the ectopic adrenal gland and because it overlaps with common symptoms, such as overweight and hypertension, in the general population (Kreitschmann-Andermahr et al., 2015). Here, we report the case of a 46-year-old male with a 15-year history of severe hypertension, facial plethora, and centripetal obesity. During treatment for herpes zoster, the patient presented with severe hypokalemia and flaccid paralysis, characteristic changes associated with CS. The serum cortisol level was elevated and baseline adrenocorticotrophic hormone (ACTH) was suppressed. After administration of the low-dose overnight dexamethasone suppression test, plasma cortisol was 38.9 μg/dL. A contrast computed tomography (CT) scan revealed normal adrenal glands and significant tumor adjacent to the right renal hilum measuring ~ 3.6 × 2.3 cm. The tumor was removed through retroperitoneoscopy, and pathological examination confirmed adrenocortical adenoma and myelolipoma metaplasia. The serum potassium level gradually became normal after surgical removal of the mass. In the current report, we have reviewed the pertinent literature and highlighted the importance of considering ectopic adrenal adenoma in the differential diagnosis of chronic CS with nonspecific symptoms.
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*Keywords:* Cushing's syndrome; Ectopic adrenal adenoma; Differential diagnosis

**Résumé**

L'adénome surrénal ectopique, responsable du syndrome de Cushing (CS), est un phénomène rare. Le diagnostic est habituellement posé des années après l'apparition de la maladie en raison du caractère insidieux que revêt la glande surrénale ectopique et parce qu'il se chevauche avec des symptômes assez banals, comme le surpoids et l'hypertension, en population générale. Nous rapportons ici le cas d'un homme de 46 ans avec 15 années d'antécédents d'hypertension artérielle sévère, de phénotype pléthorique et d'obésité centripète. À l'occasion d'un traitement pour de l'herpès, le patient a présenté une hypokaliémie et une paralysie flasque, symptômes pouvant évoquer un hypercorticisme. Le taux de cortisol plasmatique était augmenté et l'hormone adrénocorticotrophique de base (ACTH) était abaissé. Après administration de dexaméthasone pendant la nuit, le cortisol plasmatique du matin restait élevé à 38,9 μg/dL. Unscanner abdominal a révélé des glandes surrénales normales et une tumeur située dans le hile rénal droit mesurant 3,6 × 2,3 cm. La tumeur a été retirée par rétropéritonéoscopie, et l'examen pathologique a confirmé l'adénome adrénocortical associéà une métaplasie de type myélolipome. Le taux de potassium sérique est progressivement devenu normal après élimination chirurgicale de la masse. Dans cet article, nous rapportons la revue de la littérature et soulignons l'importance de considérer l'existence d'un adénome surrénalien ectopique dans le diagnostic différentiel d'un hypercorticisme devant des symptômes frustres.
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*Mots clés :* Syndrome de Cushing ; Hypercorticisme ; Adénome surrénal ectopique ; Diagnostic différentiel

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## 1. Case report

A 46-year-old man was admitted to our hospital in January 2016, because of CS presenting with hypokalemia and flaccid paralysis. During the past fifteen years, the patient gained significant weight, characterized as centripetal obesity, and developed severe hypertension with extremely high blood pressure reaching 190/130 mmHg. He took Adalat GITS 30 mg daily in the morning to control his blood pressure, that fluctuated between 120/70 and 160/100 mmHg, but did not focus on his other symptoms. Two years ago, the patient suffered from intracerebral hemorrhage, leaving a sequelae of muscle weakness in right upper extremity. Furthermore, he was diagnosed with herpes zoster based on severe pain and rash around the right intercostal skin. During anti-viral treatment, he complained of generalized weakness and fatigue predominantly involving the proximal musculature of the extremities, inability to walk and turn over, and easy bruisability of skin. Consequently, a screening test for CS was performed at the hospital, which showed a morning plasma cortisol level of 881.5 nmol/L (normal values: 171–536 nmol/L). The low-dose overnight dexamethasone suppression test revealed a plasma cortisol level of 1005.0 nmol/L. No increase in the plasma concentrations of testosterone and dehydroepiandrosterone sulfate was observed. The serum potassium level was low at 2.2 mmol/L before supplementation. The glycated hemoglobin level was 5.90%but oral glucose tolerance test disclosed impaired glucose tolerance (IGT). Metabolic alkalosis was detected via blood gas analysis. Computed tomography revealed a right para-adrenal mass and normal size of both adrenal glands.

The patient had no complaint of headaches, vision loss, polyuria, polydipsia or polyphagia. Physical examination revealed facial plethora, moon face (Fig. 1A), marked centripetal obesity, muscle atrophy of limbs, and muscle strength grade of 3+. Blood pressure was determined as 148/86 mmHg. The long-term high cortisol level was associated to osteopenia and multiple destruction of the rib cage. The patient had developed tuberculosis two years before but recovered after administration of anti-tuberculous chemotherapy. Based on the collective clinical symptoms and laboratory results, a diagnosis of CS was made.

The patient was admitted to our hospital to determine the cause of CS. The serum cortisol level showed an abnormal rhythm, with an early morning baseline ACTH level of < 1.00 pg/mL (normal values: 5.0–60.0 pg/mL). The morning plasma cortisol level was 37.2 μg/dL before and 38.9 μg/dL (normal values: 5.0–28.0 μg/dL) after the administration of dexamethasone 1.0 mg overnight. In light of the undetectable baseline morning ACTH and low dexamethasone suppression test, the patient was diagnosed with corticotropin-independent Cushing's syndrome. A contrast CT scan disclosed a significant tumor mass adjacent to the right renal hilum of ~ 3.6 × 2.3 cm in size (Fig. 2). The mass was intimately associated with the right ureter. Both adrenal glands were normal in size (Fig. 2). Laparoscopic operation facilitated removal of the adrenal tumor including right kidney because of hemorrhage of the blood vessel during tumor separation (Fig. 3).

The resection specimen consisted of a quasi-circular nodule surrounded locally by a thin film. A 3.5 × 2.5 × 2 cm mass with an irregular and lobulated shape was observed in the renal portal fibrous adipose tissue. Microscopically, tumor cells comprised a mixture of zona glomerulosa and zona fasciculata cells arranged in a nest-like, alveolar, fasciculate form. We observed no necrosis and nuclear division, and nuclear morphology was consistent. The pathological diagnosis was adrenocortical adenoma with myelolipoma metaplasia (Fig. 4). We detected a small number

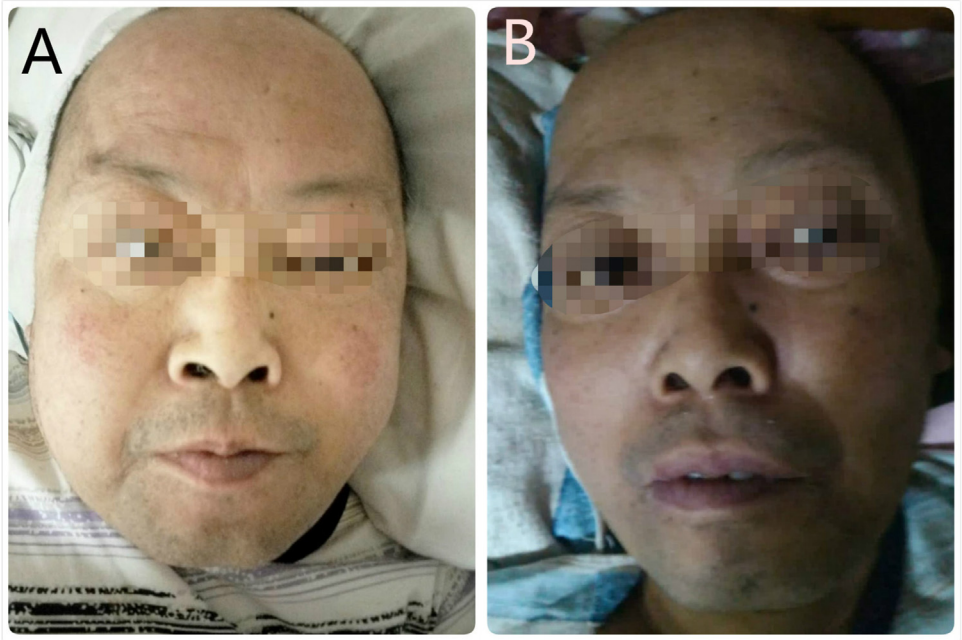


Fig. 1. Cushingoid appearance before and after surgery. A. The patient with facial plethora, moon face. B. Same patient's appearance after removal of ectopic adrenal in 6 months.

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