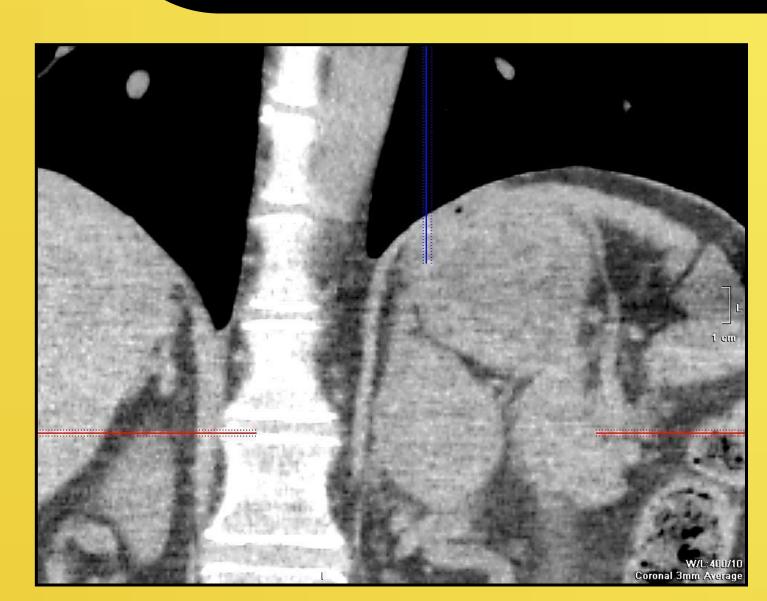
ACTH-Secreting Pheochromocytoma. Case report.

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Introduction

Cushing's syndrome has a very rare frequency with 5 cases per 1 million population. Cushing's Syndrome due to ectopic ACTH production is uncommon and due to pheochromocytoma is extremely rare.

To date, we know only few such cases.



Materials and methods

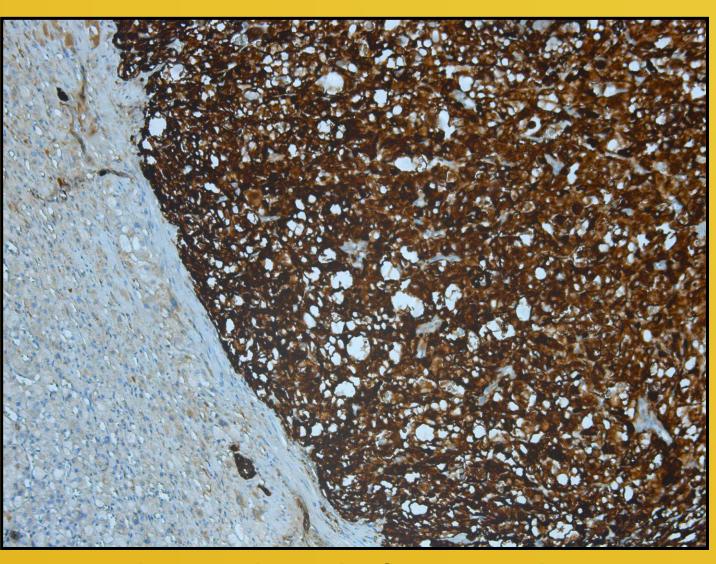
We report the clinical presentation, immunohistochemistry, imaging, histopathology, treatment of a patient with ACTH-Secreting Pheochromocytoma.





Case report

We discuss the case of a 50-year-old female who initially presented with vague, non-specific symptoms, such as general and muscle weakness, weight loss, body temperature rise, high blood pressure, increase in fasting blood glucose, in which an ACTH-secreting tumor found to be the cause of her clinical presentation. At admission: Height 168 cm, Weight 57 kg. asthenic constitution, diffusely hyperpigmented skin, darkened skin around elbows, and subcutaneous adipose tissue was insufficiently developed. Laboratory showed AM cortisol of 1488 mmol/l, PM cortisol of 1672 mmol/l, 24-hour urinary free cortisol of 3700 nmol/day, AM ACTH level of 178.7 mg/ml, PM ACTH level of 179,8 mg/ml and non-suppression of cortisol with overnight dexamethasone suppression test (1 mg and 8 mg). 24-hour urinary level of normetanephrine and metanephrine: Normetanephrine - 830 mg/day, Metanephrine - 1481 mg/day. Brain MRI showed no pathological changes. CT scan showed tumor of the left adrenal gland (2.7 x3,0x4,6 cm, density 38H). She underwent two weeks therapy by doxazozine before surgery. So, clinical and laboratory signs of Cushing syndrome and pheocromocytoma disappeared after left adrenalectomy.



Immunohistochemistry with Chromogranin A, ten

Immunohistochemistry with Synaptophysin, ten times magnification

Immunohistochemistry with ACTH, 20 times magnification

Conclusion

Despite numerous guidelines in Pheochromocytoma and Cushing syndrome, there are still diagnosis and treatment mistakes due to rarity and complexity of clinical presentation in ACTH-ectopic syndrome caused by pheocromocytoma. It is extremely difficult and important to diagnose properly before the surgical treatment to save the life of patients suffering from this disease. So, we need to improve the guidelines for diagnosis and treatment of ACTH-ectopic tumors.

Newell-Price J, Bertagna X, Grossman AB, Nieman LK. Cushing's syndrome// Lancet – 2006 - Vol 367 – P. 1605–1617

Nijhoff MF, Dekkers OM, Vleming LJ et al. ACTH-producing pheochromocytoma: Clinical considerations and concise review of the literature// European Journal of Internal Medicine – 2009 - Vol 20 – P. 682–685

Oh HC, Koh JM, Kim MS, Park JY, Shong YK, Lee KU, A case of ACTH-producing pheochromocytoma associated with pregnancy// Endocr J – 2003 - Vol 50 – P. 739-744

Kirkby-Bott J, Brunaud L, Mathonet M Ectopic et al. Hormone-Secreting Pheochromocytoma: A Francophone Observational Study// World J Surg – 2012 – Vol. 36 – P. 1382–1388