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Introduction

Pheochromocytoma is a rare, catecholamine-producing neuroendocrine tumor that arise from chromaffin cells of the adrenal medulla and the sympathetic ganglia. These tumors may rarely secrete other hormones such as ACTH.

We report a rare case of ectopic cushing syndrome due to malignant pheochromocytoma.

Case presentation

A 42-year-old man admitted to outpatient clinic with headache, irritability, vomiting, nausea, and palpitation. He had paroxysmal hypertension and urinary fractionated metanephrines were measured approximately 30 times higher than the upper limit of normal, during the hypertension attack. Abdominal computed tomography showed 43x62x78 mm mass lesion in the right adrenal gland and lymphadenopathy in the right para-aortic area (Figure 1). Endocrinological examinations demonstrated ectopic ACTH production and hypercortisolemia without overt symptoms of Cushing's syndrome. He was operated for right adrenal mass following appropriate medical preparation. Postoperative pathology revealed that malignant, metastatic pheochromocytoma. Immunohistochemistry was performed and revealed that positive staining for ACTH (Figure 2). 18F-FDG PET/CT showed that increased 18F-FDG uptake associated with 58x45 mm tumoral soft tissue mass (SUV max:6.3) in the right adrenal gland and 19 mm lymph node (SUV max: 3.8) in the retrocaval area.



Figure 1: Abdominal CT image, mass lesion in the right adrenal gland.

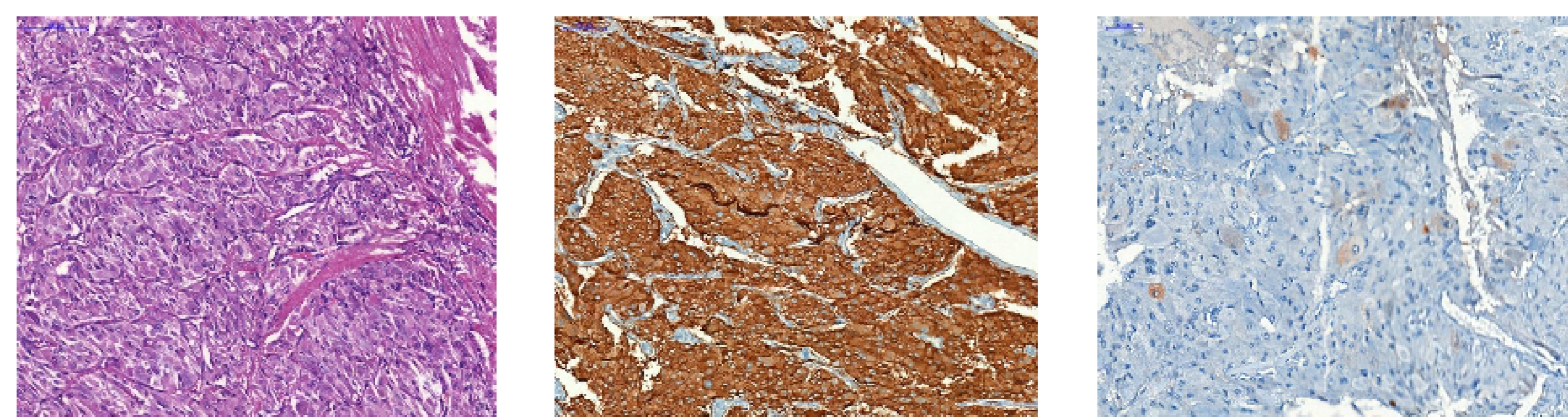
Discussion

Pheochromocytomas are functional catecholamine-secreting tumors, predominantly norepinephrine and epinephrine. These neoplasms, rarely, secrete other neurohormones such as dopamine, VIP, ACTH, β -endorphins.

Ectopic Cushing syndrome due to pheochromocytoma is a rare condition that mainly as a result of ectopic ACTH production. Ectopic hormone secretion from pheochromocytoma seems not correlated with the malignant potential of pheochromocytoma(1). However, our patient had malignant, metastatic pheochromocytoma. Pheochromocytoma is determined 3-25% of ectopic ACTH syndrome and, only 50 cases were reported in medical literature. This is named corticomedullary mix tumor. CRH-POMC derived peptides may also co-secreted.

Conclusion:

It is important to consider pheochromocytoma as a cause of ectopic Cushing syndrome. Patients with pheochromocytoma also should have tests to exclude excess cortisol secretion.



HEX20

Synaptophysin X15

ACTH X30

Figure 2: Immunohistochemical study, Positive staining for ACTH

References:

1- James Kirkby-Bott • Laurent Brunaud • Muriel Mathonet • Etienne Hamoir • Jean-Louis Kraimps • Christophe Tre sallet • Laurence Amar • Alexandre Rault • Jean-Francois Henry • Bruno Carnaille, Ectopic Hormone-Secreting Pheochromocytoma: A Francophone Observational Study., World J Surg (2012) 36:1382–1388.

