

Association of pheochromocytoma and preclinical cushing's syndrome

Abdelkefi Ch, Haouat E, Hbaili N, Ben Salem L, Ben Slama C
National Institute of Nutrition – Tunis- Tunisia

Introduction

Pheochromocytoma and Cushing syndrome are two uncommon endocrine conditions. The association of these two conditions is very rare and not well elucidated.

We report an association of a pheochromocytoma with an ACTH-dependant Cushing syndrome.

Case report

A 32 years old female patient was admitted in the endocrine department for paroxysmal hypertension diagnosed in the immediate post partum period. She reported sweating and palpitation crises occurring with hypertensive peaks. Her blood pressure on treatment (calcic inhibitor and central antihypertensive) was 95/70. Clinical examination didn't reveal any evidence for Cushing syndrome. Laboratory findings showed hypokalemia at 3.8 meq/l with a hyperkaliuresis.

CT-scan revealed a 34x34mm homogenous right adrenal mass. Twenty four hours urinary metanephrines were 3 folds the upper limit of the normal, confirming pheochromocytoma.

Cortisol after low dose dexamethasone suppressing test (LDDST) was 60 nmol/l confirming the diagnosis of Cushing syndrome. cortisol was suppressed by high dose dexamethasone test (basal cortisol: 236.1nmol/l and cortisol after high dose dexamethasone test : 105.6nmol/l) . ACTH was 39 ng/l. Pituitary MRI was normal. The patient had an endoscopic adrenalectomy with no complications. Histology concluded to pheochromocytoma without signs of malignancy. Immunohistochemistry was not performed.

After surgery, hypertension and hypokaliemia disappeared. Urinary metanephrines became normal and cortisol was suppressed by LDDST (cortidol after LDDST: 15.6 nmol/l).

Discussion

The coexistence of pheocromocytoma and Cushing's syndrome has already been reported. Few cases do exist in the literature. It is caused by an adrenal tumor secreting catecholamines and ACTH or precursors of ACTH resulting in a pheochromocytoma with an ACTH dependant paraneoplastic Cushing Syndrome. Usually an adrenal hyperplasia induced by the ACTH excess is reported on CT scan and on histology.

The Cushing syndrome in these situations is usually subclinical, diagnosed on biological assessment, ACTH is also slightly elevated and cortisol is usually not suppressed by the high dose dexamethasone suppressing test (HDDST).

The particularity of our case, is that cortisol was suppressed by the HDDST. It was necessary to rule out a Cushing disease associated with pheochromocytoma by performing at least a pituitary MRI.

Conclusion

This case report illustrates a very rare cause of Subclinical Cushing syndrome: a pheochromocytoma with a paraneoplastic ACTH secretion.

References:

Guzin : pheocromocytoma combined with subclinical Cushing's syndrome and pituitary microadenoma

Erem : Pheocromocytoma combined with pre-clinical Cushing's syndrome in the same adrenal gland.

