Title:

A rapid thymic carcinoma in a patient with multiple endocrine neoplasia type 1

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Abstract;

Introduction

Thymic neuroendocrine (NE) tumors associated with multiple endocrine neoplasia type 1 (MEN-1) are rare, variably documented in 1-8% cases. They are malignant and aggressive tumors and form a major cause of mortality in MEN-1.
We report a case of thymic NE carcinoma developing rapidly after parathyroidectomy in a MEN-1 patient.

Case

A 45 years-old man was allowed for headaches, the biology showed a hypercalcemia, a hypophosphoraemia and high PTH.The imaging revealed polar parathyroidal adenoma with hypophyseal adenoma with rates of prolactinemia = 1768 MU/ml. The abdominal MRI - Pet Scan - FDG and the scintigraphy in the ocréotide allowed to bring to light a metabolic activity of the mass of the thymic cavity with a necrotic and atypical aspect without distant metastases, 5 pancreatic nodules and bilateral adrenal nodules without arguments in favour of a pheochromocytoma.

The patient has benefited from the resection of a retro-tracheal parathyroidal adenoma, from a malignant thymoma of type invasive P3 the pericardium and the mediastinal fat completed by a mediastinalradiotherapy and hypophyseal adenomectomy.

The search of MEN 1 showed itself positive.

Conclusion

Thymic NE tumors in MEN-1 are commoner in males and smokers and are almost always hormonally inactive and diagnosed incidentally .They are malignant, aggressive tumors and are widely invasive and metastatic at presentation (usually to bone).They are never the presenting feature of MEN-1 and almost always occur after hyperparathyroidism, providing an opportunity for prophylaxis for these tumors at the time of parathyroid surgery.

Thymic NE tumors present later, usually 15-20 years after. In contrast, our patient had a rapid presentation of thymic NE carcinoma.

Keywords: Multiple endocrine neoplasia type 1, thymic neuroendocrine carcinoma.