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(CASE REPORT)

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A rare case of Cushing syndrome ectopic-ACTH: A case report

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Abstract

Malignant tumors, nevertheless, have been associated with extremely high circulating ACTH and cortisol levels, and short duration of symptoms of Cushing syndrome (CS) besides atypical clinical phenotype, when compared with pituitary-dependent Cushing. A 33-year-old woman with a history of insulin-induced diabetes admitted to our department for suspicion of ACTH-dependent Cushing's syndrome held in front of : a Cushing's syndrome clinic, biological Cushing's syndrome, CLU : 2894ug/24h, minute braking : negative, cortisolemia 32.1ug/dl, ACTH : 232ng/l, CT scan of the adrenal glands in favor of moderate adrenal hyperplasia. MRI Pituitary without anomaly, strong negative braking. An Octreoscan objectified the presence of two thyroid nodules of hypodense generation moderately fixing the radiotracer, measuring respectively 1.9*1.2cm on the right and 1.5*1.1cm on the left. The focus intensely fixes the retrotracheal radiotracer and the right paravertebral retrotracer. Tumor markers are negative. The patient was operated on for anopathy : thyroid nodular dystrophy, reactive lymphadenitis, a review of the anopathy with iminohistochemistry is requested results in progress. The patient comes to our training 1 month post-operative, the patient reports a regular cycle return, the clinical examination shows a disappearance of purple stretch marks and bruises. Control octreoscan shows the absence of signs in favor of a neuroendocrine tumor at the pleuro-pulmonary, digestive or thyroid level. The nodule resected to objective the presence of anti-ACTH antibodies in immunohistochemistry. . Identification of the source of ACTH can be challenging, as sometimes the primary lesion is not identified even after prolonged and repeated follow-up.

Keywords: Anti-ACTH antibodies; Cushing syndrome; Ectopic-ACTH; Nodules; Immunohistochemistry

1. Introduction

Diagnosis of ACTH ectopic secretion involves two basic steps that cannot be omitted to prevent misdiagno confirmation of hypercortisolism and determination of its etiology (differential diagnosis) [1] . Identification of the primary tumor responsible for the ectopic ACTH production may be troublesome. CT, PET-FDG, and octreoscan have equal, but low sensitivities for this purpose. They must complement each other in difficult cases. A variety of factors may influence the ability of FDG-PET to locate a tumor. Increased metabolic rate and glucose transport through tumor cell membranes are necessary for increased up take of FDG [2]. Development of very simple methods for measuring the ACTH precursors has demonstrated that they are released into the circulation in normal subjects and that the levels are markedly elevated in non-pituitary tumours, suggesting that they are the major circulating forms in the ectopic ACTH syndrome [3] .

2. Case presentation

A 33-year-old woman with a history of insulin-induced diabetes admitted to our department for suspicion of ACTH-dependent Cushing's syndrome held in front of : a Cushing's syndrome clinic, biological Cushing's syndrome, CLU : 2894ug/24h, minute braking : negative, cortisolemia 32.1ug/dl, ACTH : 232ng/l, CT scan of the adrenal glands in favor

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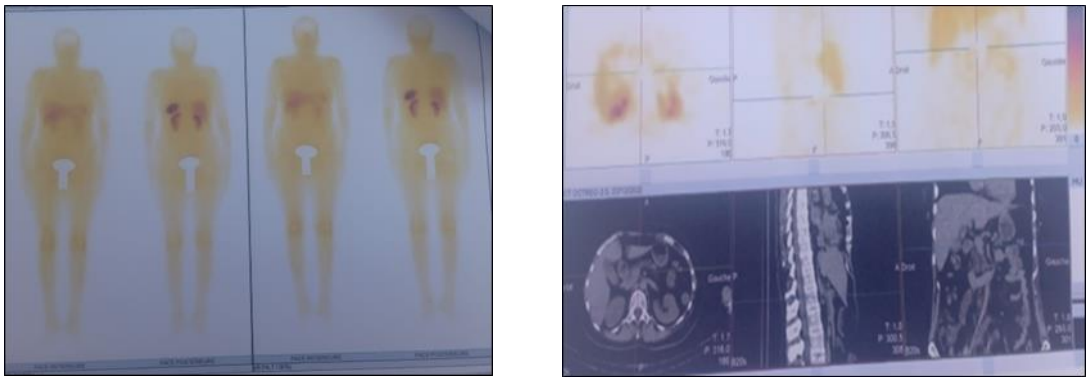

Figure 1 Octreoscan the presence of two hypodense bilateral thyroid nodules

3. Discussion

Most primary endocrine tumors responsible for Ectopic ACTH syndrome (EAS) are located in the chest. Among all ACTH-secreting thoracic tumors, the most common, in a modern endocrine patient recruitment, are welldifferentiated NETs located in the bronchi (formerly called 'carcinoids') and these account for 20% to 40% of all cases of EAS in recent series . They can recur, especially after initial resection without systematic lymphadenectomy . 'Tumorlets' or diffuse idiopathic pulmonary neuroendocrine cell hyperplasia, a precursor to carcinoid tumors and tumorlets, represent a particular bronchial NET type, being small These are mainly located in the chest and can mimic lung metastases. Thymic NETs are also an important cause of EAS due to thoracic tumors accounting for 5% to 10% of EATs, depending on the series. Numerous types of endocrine and non-endocrine tumors acquire the ability to secrete substances that are not usually produced by the normal tissue from which the tumor is originated [4] .Various solid tumors, mainly of neuroendocrine origin, are as well recognized as ACTH-secreting ones, causing ectopic-ACTH secretion [5] . Either benign or malignant tumors may be the cause of ectopic ACTH syndrome. ACTH- -dependent Cushing syndrome must invest all available resources in shortening ACTH production site location. Several distinct locations, and association with others neuroendocrine tumors warrants such a strenuous effort. Ectopic production of ACTH by chemodectomas is a rare event. As far as we know, there is only one case reported in literature associating a neck paraganglioma with CS [6]. Another patient with CS has been reported, in whom resection of a chemodectoma following bilateral adrenalectomy resulted in decreased ACTH levels. Nonetheless, there was no evidence of cyclic ACTH production [7].

4. Conclusion

ACTH-dependent Cushing syndrome due to ectopic ACTH production most of times is difficult to manage. The identification of the source of ACTH may take many years until final diagnosis. ACTH-dependent Cushing syndrome due to ectopic ACTH production most of times is difficult to manage. The identification of the source of ACTH may take many years until final diagnosis.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

The authors declare no conflict of interests.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subject by any of the authors.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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