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Case Report
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Large Neuroendocrine Tumor in a Patient with Acromegaly Case Report

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Abstract

The case of a patient with a rare malignancy whose incidence ranges from 0.2 to 2/100,000 individuals. The increase in incidence probably showed increasing changes in diagnosis. Approximately 25% of these tumors grow in the lungs and represent 2% of all lung tumors. Of all bronchial NETs, only 5% are associated with Multiple type 1 Endocrine Neoplasms (MEN-1). According to the current WHO classification of 2015, neuroendocrine lung cancer should be classified into typical carcinoids, atypical carcinoids, large cell neuroendocrine carcinoma or small cell carcinoma, the classification being based on histological morphology. The commonly associated endocrine syndromes are inadequate secretion of the natriuretic hormone, Cushing's syndrome and, rarely, acromegaly. We report a case of a male patient with a large neuroendocrine tumor associated with a pituitary macroadenoma. Laboratory tests on admission were changed, GH of 56.5 ng/mL (Ref <5 ng/mL), IGF-1 of 1304.4 ng/mL (Ref- 96.4 - 227.8 ng/mL), chromogranin 4.89 (Ref 3.00), b-HCG-negative. The patient was initially submitted to thoracic surgery and subsequently underwent neurological surgery in another center, so that he was accompanied by professionals from various areas, making it increasingly evident the need for hospitals to have centers for the study of these neuroendocrine tumors.

Keywords: Acromegaly; Lung cancer; Adenoma pituitary; MEN-1

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Case Presentation

A 31-year-old man with acromegaly diagnosed on admission examination with enlarged image of the cardiac area on chest X-ray. A transthoracic echocardiogram noted a giant mass beyond the heart. There have been allegations of pulsatile parieto-occipital headache associated with nausea and vomiting and back pain. Over the past 10 years, he has experienced pain and paresthesias in the extremities, and for the past five years, he has seen vision problems with hemianopsia (partialor complete loss of vision in one or both eye halves) in the right eye and reduced visual field in the left eye. In addition, he refers to malaise, recent and late memory reduction and libido. In our pulmonology department, laboratory tests were performed and are shown in Table 1.

A transthoracic echocardiogram marks a large giant intrapericardial mass with right atrial compression with 30 mmHg PSAP. Cranial magnetic resonance imaging diagnosed a pituitary mass with infra and suprasellar extension of 3.4 cm × 2.5 cm × 3.2 cm, characterizing a macroadenoma. Chest computed tomography showed a large mass with a huge cardiac deviation. Bronchoscopy confirms massive extrinsic airway compression with an exophytic mass in the intermediate bronchus. A somatostatin scintigraphy examination showed hypercaptation in the topography of the turcic saddle, in the antero-inferior portion of the right hemithorax and in the inguinal regions, being more pronounced on the right (Figure 1).

Sternotomy was performed subsequently (Figure 2 and 3), first to ensure that there was no cardiac invasion that might require cardiac bypass. Since the previous echocardiography suspected cardiac involvement, an intraoperative trans-esophageal echocardiogram was performed confirming that the previous cardiac image was due to the Eustachian valve (Small crescentic valve partially

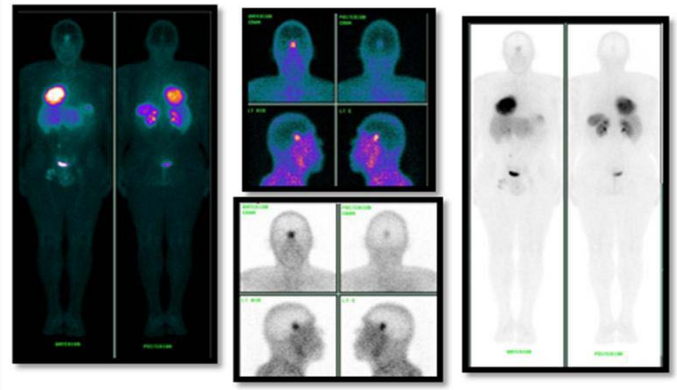

Figure 1: Somatostatin scintigraphy.

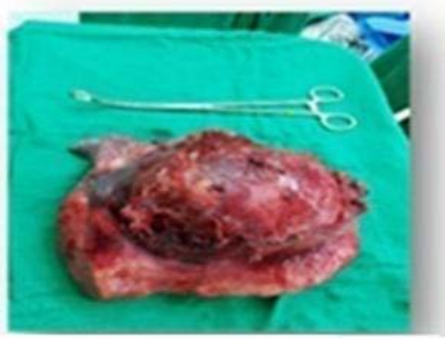

Figure 2: Lung TNE.

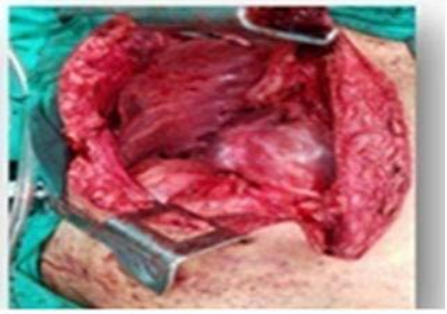

Figure 3: Surgical bed.

enclosing the inferior vena cava hole in the right atrium). After opening the pericardium to exclude cardiac invasion, a combined right anterior thoracotomy was performed. Due to the topography of the lesion, a right intrapericardial pneumonectomy was performed with the entire right pericardial sac in there section of the block. The postoperative period was uneventful and the patient was discharged shortly thereafter. The biochemical test confirms the normal levels of returned hormones (Table 1). In agreement with the other center treating this patient, it was planned to perform lung Neuroendocrine Tumor (TNE) surgery initially and to evaluate the pituitary adenoma at another time.

Discussion

Clinical manifestations of acromegaly in patients with ectopic GHRH syndrome are indistinguishable from those of any GH secreting pituitary adenoma, and regardless of the cause, serum GH

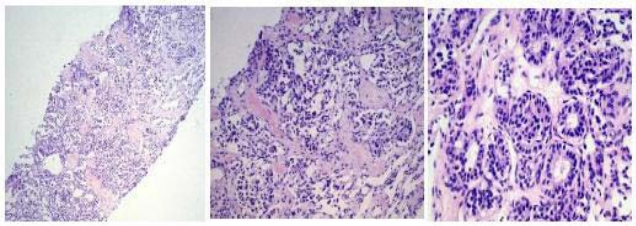

Figure 4: Photobiography of lung neuroendocrine tumor. Panoramic photo, 20x and 40x magnification respectively.

and IGF-1 levels are invariably elevated and GH levels are not [1]. Can suppress (<1 ng/mL) during TOTG in all forms of acromegaly. Of all, plasma GHRH is the most accurate test for the diagnosis of ectopic GHRH, causing acromegaly, but it comes at a cost. In our patient, bronchial carcinoid was visualized by chest CT and octreoscan [2]. However, specificity is limited because scintigraphy is positive in many other tumors and not all carcinoid tumors expressing somatostatin receptors by immunohistochemistry are positive for octreoscan [3].

Laboratory tests at admission were altered as shown in Table 1, GH of 56.5 ng/mL (Ref <5 ng/mL), IGF-1 of 1304.4 ng/mL (Ref-96.4 - 227.8 ng/mL), Chromogranin of 4.89 (Ref 3.00), b-HCG-negative. Hyperparathyroidism was removed and genetic examination was not performed. Primary hyperparathyroidism, present in 90% to 97% of patients with MEN-1 syndrome, is usually the first manifestation of this syndrome, but was not present in this case [4].

Thus, three other issues need to be considered. At first, the coexisting carcinoid tumor and the possible pituitary adenoma alerted us to the possibility of MEN-1. Secondly, bronchial carcinoid may have metastasis to the pituitary gland. And lastly, the acromegaly may have been due to the pituitary tumor producing excessive amounts of GH, and its self-infarction leads to normalization of IGF-1, a normal GH response following a Glucose Tolerance Test (TOTG) and shrinkage of the tumor on subsequent magnetic resonance imaging. After the surgical procedure has normalized IGF1, and subsequent imaging study confirms the reduction or resolution of the pituitary lesion after lobectomy, the first possibility of a MEN-1 is unlikely. Although there is no evidence of histopathological examination of the pituitary tumor, again, the disappearance or reduction of the lesion after lobectomy also makes the possibility of metastasis unlikely. In relation to the third possibility, it cannot be proved or refracted in the absence of a plasma GHRH level and tumor histological type.

Table 1: Pre and post markers for acromegaly.

	Preoperative examination	Postoperative (3 months)	Postoperative Adenoma (3 months)	Reference values
Blood Glucose (mg%)	125 mg%			<99 mg%
25 OH vitamin D (ng/mL)	21.8 ng/mL	40.3 ng/mL		30-100 ng/mL
Testosterone (ng/dl)	<20 ng/dl	282 ng/dl		262-1,593 ng/dl
Basal Cortisol (mcg/dL)	8.3 mcg/dL	17.8 mcg/dL		5-25 mcg/dL
IGF1 (Somatomedine C) (ng/mL)	1304.4 ng/mL	325.9 ng/mL		96.4-227.8 ng/mL
GH (ng/mL)	56.5 ng/mL	1.5 ng/mL	0.58 ng/mL	<5 ng/mL
Prolactina (ng/mL)	109 ng/mL	<0.5 ng/mL		2.5-17 ng/mL
TSH (mU/L)	0.7 mU/L			0.4-4.0 mU/L
Free T4 (ng/dL)	1.11 ng/dL			0.8-1.76 ng/dL
LH (mUI/mL)	4.2 mUI/mL			0.8-7.6 mUI/mL

GH: Growth Hormone; IGF1: Type 1 Insulin Growth Factor; TSH: Thyroid Stimulating Hormone; LH: Luteotrophic Hormonium

However, it is necessary to follow the patient closely to get the final answer. In this case, normalization of the exams has already been shown in the immediate postoperative period, as shown in the table above (Table 1). However, the patient was already using somatostatin analogs.

Surgical resection is the gold standard in the treatment of bronchial carcinoids because it offers the best chance of cure for the patient. The prognosis for subsequent resection of a typical carcinoid is excellent, with reported 5-year survival rates of 87% to 100%. While for a typical carcinoid, 5-year survival of 30% to 95% has been reported. Chemotherapy and radiotherapy are generally not effective. Long-term somatostatin analogues provide an effective option to control symptoms and, according to some studies, may also slow tumor progression [5-15].

The postoperative period was uneventful with the simple removal of the chest tube on October 19th, 2018 and discharge on October 22nd, 2018. The histopathology also confirmed that it was a carcinoid tumor (Figure 4), later confirmed by Immunohistochemistry.

With the favorable evolution of the case, the patient after discharge from the surgery was referred to and continued his clinical follow-up with endocrinology, in order to be submitted to pituitary surgery, however, the patient only visited the clinic on November 22nd, 2018, when hypovitaminosis D was observed, colecalciferol was prescribed and some preoperative exams were requested. He reported that after removal of the lung tumor, he showed improvement in symptoms, chest pain, dyspnea, and limb edema. Reports use of medication for hypertension, diabetes. It also shows improvement in the erection, after the use of octreotide and cabergoline started in September 2018.

Due to a sinus disease, pituitary surgery was postponed and underwent only transsphenoidal surgery, confirming the adenoma and MEN-1 in April 2019.

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