Cervical paraganglioma mimicking a thyroid nodule with recurrent nerve involvement☆

Paraganglioma cervical simulando nódulo tiroideo con afectación del nervio recurrente

Diagnosis of recurrent laryngeal nerve paralysis associated with an ipsilateral thyroid nodule is highly suggestive of thyroid cancer. We report a case with these initial findings in which a much more uncommon condition was diagnosed: a solitary, sporadic, non-functioning cervical paraganglioma.

The patient was a 38-year-old woman with an unremarkable family history. Her clinical history only included bronchial asthma for which drug treatment was not currently being administered. She had experienced no asthma attacks in recent years.

The patient reported dysphonia for approximately 3 months, and was therefore initially seen at the otolaryngology outpatient clinic. Laryngoscopy revealed left vocal cord paralysis. A computed tomography (CT) scan showed a nodular lesion, approximately 3 cm in size, apparently dependent on the most caudal portion of left thyroid lobe, with posterior and descending extension almost to the sternal manubrium. There were signs of left recurrent nerve paralysis and dilation of the ipsilateral laryngeal ventricle. Thyroid function was normal. Ultrasound examination with fine needle aspiration (FNA) for cytological study of the nodule was requested. Ultrasonography revealed a single 33 mm, hypoechoic thyroid nodule in a very inferior and posterior position at the left thyroid lobe. FNA was performed with difficulty because the nodule was not readily accessible, and the material collected was insufficient for diagnosis.

Because of the suspected signs of the nodule and the difficulty in performing FNA, a repeat FNA was not done, and surgical lesion resection was performed instead. A left hemithyroidectomy was performed, and a nodule adherent to the pharyngolaryngeal muscles that had destroyed the left recurrent nerve was found and resected. Perioperative examination of the surgical specimen revealed suspected medullary thyroid cancer. A total thyroidectomy and excision of the central and left cervical lymph nodes were therefore performed. The final pathological diagnosis, once the tissue was shown to be neuroendocrine in nature but negative for calcitonin at immunohistochemistry, was a 2.7 cm paraganglioma adherent to the lower pole of the left thyroid lobe with no signs of malignancy. Thyroid parenchyma was normal, and the lymph nodes were negative for malignant cells.

Subsequent repeated measurements of fractionated metanephrines and catecholamines in acidified 24-h urine were normal. A metaiodobenzylguanidine (MIBG) total body scan was negative, and a CT scan of the chest and abdomen ruled out other paragangliomas. The patient was therefore diagnosed with a solitary benign cervical paraganglioma, probably non-functioning based on the absence of adrenergic symptoms during tumor manipulation.

Despite the absence of a family history, a genetic study was performed to rule out mutations in succinate dehydrogenase (SDH) genes, initially SDHA and SDHC and, after negative testing, SDHB and a genetic study for von Hippel–Lindau disease. All of these tests were negative.

The patient is currently receiving replacement therapy for postoperative hypothyroidism. Vocal cord paralysis has been well compensated and remains asymptomatic.

Paragangliomas are neuroendocrine tumors arising from the extra-adrenal chromaffin tissue of the autonomic nervous system that may occur in the base of the skull, the neck, the chest, and the abdomen. Cervical tumors account for 0.012% of all human tumors and for 0.6% of head and neck tumors.[1] Most paragangliomas are highly vascularized, benign, and non-functioning. Malignancy is established based on the presence of lymphatic or hematogenous dissemination, rather than on histological findings in the specimen.[2] Surgical resection is the treatment of choice, and associated chemotherapy or radiotherapy may be performed in malignant cases.[2] It is estimated that up to 30% of cervical paragangliomas may be familial.[1] Genetic study is therefore recommended, because a positive study involves a risk of developing new tumors and may have implications for the screening of relatives. The genetic changes most closely related to the development of cervical paragangliomas are those occurring in SHD genes, particularly SDHD and SDHC,[3] but a significant number of SDHB mutations have also been documented in the Spanish population.[1] They may also be part of von Hippel-Lindau disease and neurofibromatosis type 1. Genetic study of these conditions may therefore be considered if suggested by other clinical data.

To sum up, cervical paragangliomas are uncommon tumors that should be considered in differential diagnosis of neck masses.

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Diabetes mellitus onset in young patient: Type 1 diabetes?☆

Debut de diabetes mellitus en paciente joven: ¿diabetes tipo 1?

Symptomatic diabetes as a manifestation of another underlying condition is uncommon, and usually due to the use of drugs with hyperglycemic potential or to neoplastic processes in the pancreatic area. Among drugs, special mention should be made of high-dose steroids and, more recently and in young patients, atypical neuroleptics.[1] The onset of cardinal symptoms of diabetes mellitus (DM) in a patient under 40 years of age, with no other organ-specific clinical signs or concomitant treatment suggests autoimmune diabetes (type 1A) as the first diagnostic possibility, while in elderly patients it should rule out an underlying neoplastic condition. We report the case of an apparent onset of type 1 DM in which subsequent evaluation showed an unexpected, reversible etiological diagnosis.

A 38-year-old male patient with unremarkable personal or family history was referred from the emergency room for hyperglycemia with ketosis. The patient reported urinary frequency and severe polydipsia for the previous three weeks, and the loss of 12 kg in the previous year with no decreased appetite or other symptoms. The results of tests performed in the emergency room showed blood glucose 340 mg/dL, ketone bodies in urine 50 mg/dL, and pH 7.4, weight 56.5 kg (BMI 17 kg/m^2), blood pressure 145/95 mmHg, and heart rate 100 beats per minute, with no other physical examination findings. Basal-bolus insulin therapy (glargine and aspart) and diabetes education were started, and good glycemic control was achieved. Glycosylated hemoglobin at onset was 13.4%, with negative anti-IA2 (insulinoma antigen 2) and anti-GAD (glutamic acid decarboxylase) antibodies, and a basal C-reactive peptide level of 2.2 ng/mL. Diagnosis of type 1A DM was therefore discarded, and additional tests were performed to rule out secondary causes.

Levels of thyroid hormones, cortisol, calcitonin, calcium, and phosphorus were normal. Abdominal ultrasound revealed a 7-cm right adrenal mass, which magnetic resonance imaging confirmed to be a heterogeneous tumor with a necrotic center (Fig. 1). The results of measurements of catecholamines and their metabolites in 24-h urine were as follows: norepinephrine 767 μg/24 h (normal [N]: 12.1–85.5), epinephrine 270 μg/24 h (N: 1.7–22.4), dopamine 461 μg/24 h (N: 0–498), normetanephrines 3245 μg/24 h (N: 88–444), and metanephrines 2299 μg/24 h (N: 52–341). After preoperative preparation with phenoxybenzamine (no beta-blockers were required), laparoscopic adrenalectomy was successfully performed at four weeks. A pathological study confirmed a 7.5 cm × 5 cm × 5 cm pheochromocytoma, 138 g in weight, without capsular invasion. Insulin requirements dramatically decreased after surgery, and hypoglycemic therapy was not required after a few days. At the time of writing, the patient remains asymptomatic, euglycemic, and with normal blood pressure values He has recovered his normal weight and his hormone levels have remained normal.

Pheochromocytoma is a catecholamine-secreting tumor derived from the enterochromaffin cells of the adrenal medulla. It is an uncommon tumor, diagnosed in approximately 1–2 cases per 100,000 inhabitants a year,[2] although its prevalence is greater (0.05–0.1%) in autopsy studies.[3,4] Clinical presentation widely ranges from asymptomatic patients to severe hypertensive crises.[5] Although the classical triad of palpitations, headache, and sweating is highly specific for diagnosis, most patients have sustained or have paroxysmal arterial hypertension[5] with

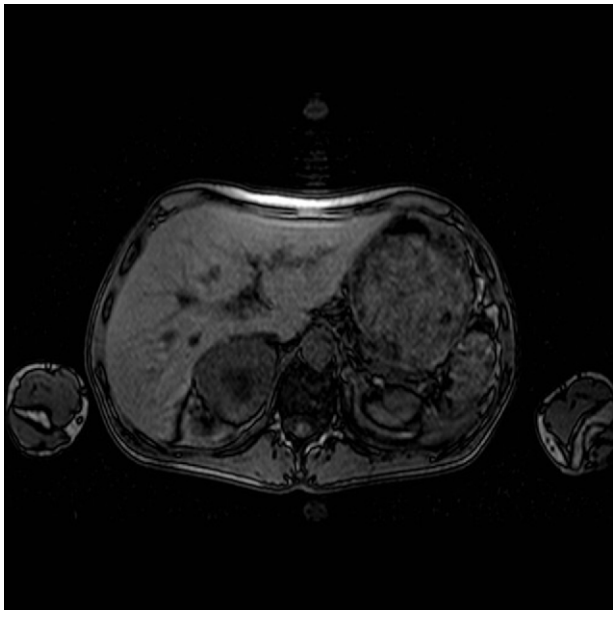

Figure 1 Heterogeneous lesion with necrotic center in the right adrenal gland.

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