ABSTRACT

Mediastinal Paragangliomas are low incidence tumors that arise from neural crest. Its

diferential diagnosis include several malignant diseases, so its correct characterization is

mandatory for an adecuate therapeutic management. 111In-Octreotide scintigraphy/SPECT-CT

can detect Paraganglioma, mainly in those cases of atypical or unsuspected location, and may

have a significant role in follow-up of those patients diagnosed with Multifocal and Familiar

 Paraganglioma. Here, we report a rare case of middle mediastinal Paraganglioma detected by

111In-Octreotide scan and SPECT-CT, which allowed an optimal treatment.

INTRODUCTION

Paragangliomas are low incidence neuroendocrine tumors that express somatostatin

receptors, being type 2 the most common of them 1.2. They arise from the neural crest of

the autonomic nervous system and are usually benign slow growing lesions, which can

produce compressive clinic or neurological dysfunction due to involvement of adjacent

nerves. Its most common location is head and neck, being the carotid glomus or carotid

Chemodectoma the most frequent of all its forms 3,4.

Diagnosis is based on structural imaging tests such as magnetic resonance imaging (MRI),

computed tomography (CT) or angiography and functional imaging tests, as

111In-Pentetreotide scintigraphy. One of the main diagnostic indications of Nuclear

Medicine tests is the suspicion of paragangliomas in areas of difficult biopsy access and/or

in cases of atypical locations, which require differential diagnoses with other pathologies1.

Here we report a case of an extremely rare middle mediastinal paraganglioma, detected by

scintigraphy and SPECT-CT with 111In-Octreotide in a patient diagnosed of multifocal and

family history of paraganglioma.

CASE

A 54 year old male was diagnosed in 1997 of family and multifocal paraganglioma

(bilateral carotid and right jugulotympanic paraganglioma). He underwent surgery and all

paraganglioma were excissed. Afterwards, he stayed asymptomatic with no evidence of

residual disease or recurrence by structural techniques and octreotide and being

discharged in 2005.

In 2013 he returns to our Hospital for a follow up consultation. Otoscopy and analytical

study showed no abnormalities and 111In-Octreotide scintigraphy was requested to

evaluate rest or recurrence of paraganglioma. The study showed weak uptake in the left

jugular region, what led to the suspicion of tumor recurrence, and review was scheduled

within a year.

New scintigraphic control was performed in October 2014, showing the previously

described weak focal uptake and the appearence of a new focus in anterior mediastinum

(IMAGE 1). Thoracic SPECT-CT fusion images located the mediastinal focus adjacent to

aorta and pulmonary trunk (IMAGE 2). Given this finding, contrast enhanced CT

scan and an angiography were performed. CT scan reported a nodule of 15 x 18 mm in

aortopulmonary window, adjacent to the aortic root, in contact with the left coronary sinus.

The angiography showed nodule irrigation by septal coronary branch. No MRI was

performed because of multiple ferromagnetic plates in both lower limbs, due to an accident

suffered by the patient 20 years ago.

Given these findings, surgical treatment of the mediastinal lesion was planned. Surgery

was carried out by median sternotomy and extracorporeal circulation. The pathological

examination of the surgical specimen showed para-aortic low-grade tumor, positive for

chromogranin and synaptophysin, cytokeratin AE1-AE3 negative and Ki67 lower than 5%,

compatible with paraganglioma (FIGURE 1). Currently, the patient is tumor free, under

clinical monitoring.

DISCUSSION

Mediastinal paragangliomas are extremely rare. They are most commonly located in the

posterior mediastinum, with its origin in paraspinal ganglia, while those located in the

middle mediastinum arise from the para-aortic lymph node chains. Its low frequency forces

to establish a differential diagnosis with other pathologies such as thymoma, thymic

carcinoma, metastases or angiosarcoma7. However, these pathologies do not express

somatostatin receptors, so 111In-Pentetreotide scintigraphy exclude them. 5.6.

The first line diagnosis of paraganglioma is the study of biochemical parameters and

structural imaging tests (CT, MRI). Scintigraphy and SPECT/CT with 111In-Octreotide

have good levels of sensitivity (93%) and specificity (86,5 %) for the detection of head and

neck paragangliomas 3,8. FDG PET studies are especially useful in cases with negative

scintigraphy and positive mutation for succinate dehydrogenase gene (SDHX) 9. In this

case, the patient showed negative biochemical tests and SPECT/CT with 111In-Octreotide

identified the para-aortic lesion accurately, allowing its proper surgical excision.

In conclusion, we report the case of a 54 years old male with multifocal and family history

of paraganglioma 17 years ago, who currently presented an extremely rare asympyomatic

middle mediastinal paraganglioma, diagnosed by planar and SPECT/CT with 111in-

Pentetreotide scan.

This procedure proved to be a helpful diagnostic method because of its potential to explore

full body, so It allows us to locate unsuspected and atypical location paragangliomas. This

finding suggests that patients diagnosed of multifocal head and neck paraganglioma

should undergo periodical follow-up with 111In-Pentetreotide scan to detect unsuspected

paraganglioma.

Disclosure: The authors state no conflict of interests.

REFERENCES

1. C. Castillo-Berrio, M. Castrillón, F. Zelaya et al. SPECT-TC con 111In-octreotide en paragangliomas de cabeza y cuello. Rev Esp Med Nucl Imagen Mol. 2015 Sep-Oct;34(5):321-4. doi: 10.1016/j.remn.2015.02.007. Epub 2015 Apr 6.

2. Matthias Schmidt, Eva Fischer, Markus Dietlein et al. Clinical value of somatostatin receptor imaging in patients with suspected head and neck paragangliomas. Eur J Nucl Med Mol Imaging. 2002 Dec 29(12):1571-80. Epub 2002 Sep 21.

3. Tamayo P, Ruano R, Muñoz A. Diagnóstico y control evolutivo de los paragangliomas de cabeza y cuello. Aportaciones de la medicina nuclear. Acta Otorrinolaringol Esp. 2009;60:68–75.

4. Feijoo C, Carranza JM, Rivera MI, et al. Tumores del cuerpo carotídeo.Experiencia en 22años y protocolo de seguimiento y despistaje familiar. Angiologia. 2012;64:155–60.

5. G. Bano, D. Sennik, M. Kenchaiah, et al. A case of co-existing paraganglioma and thymoma. Springerplus. 2015 Oct 21;4:632. doi: 10.1186/s40064-015-1269-z. eCollection 2015.

6. Christopher K. Mehta MD, Colin T, et al. Rare Middle Mediastinal Paraganglioma Mimicking Metastatic Neuroendocrine Tumor. Ann Thorac Surg. 2015 Aug;100(2):702-5. doi: 10.1016/j.athoracsur.2014.09.068.

7-Shibahara J, Goto A, Niki T, et al. Primary pulmonary paraganglioma: report of a functioning case with inmunohistochemical and ultrastructural study. Am J Surg Pathol 2004;28:825-9.

8-Bustillo A, Telisci F, Weed D, et al. Octreotide scintigraphy in the head and neck. Laryngoscope. 2004; 114: 434-40.

9-Darr R, Lenders JWM, Hofbauer LC et al. Pheochromocytoma-update on disease management. Ther Adv Endocrinol Metab. 2012;3:11-26.

10-William D. Travis Elisabeth Brambilla, Allen P. Burke, Alexander Marx, Andrew G. Nicholson (Eds.): WHO Classification of Tumors of the Lung, Pleura, Thymus and Heart. IARC: Lyon, 2015.

LEGENDS

LEGEND FIGURE 1 AND FIGURE 2 Aortic Paraganglioma. 111In-Octreotide planar scintigraphy and SPECT-CT images show weak focal uptake in mediastinum, adjacent to ascendant Aorta. Histologic study was recommended.

LEGEND FIGURE 3 Its histology consists of two main celular types: epitelial (3**A**) and peripherical sustentacular cells; Both involved in a highly vascularized Stroma. The presence of citological atypia and celular pleomorfism (3**B**) are not a sign of agressiveness, as this is determined by the presence of necrosis, mitosis and capsular invasión. Its inmunohistochemical features are: Sinaptophisine/cromogranin positive (major cells) (3**C**) and s-100 sustentacular cells (3**D**) with a low (Ki-67) proliferation index. Citocheratin (actin and desmin) expression were negative. Those findings lead us to the diagnosis of aortic paraganglioma.10