Clinical note

Pre-operative $^{68}$Ga-DOTANOC somatostatin receptor PET/CT imaging demonstrating multiple synchronous lesions in a patient with head and neck paraganglioma

N. Naswa, S. Karunanithi, P. Sharma, R. Soundararajan, C. Bal, R. Kumar*

Department of Nuclear Medicine, All India Institute of Medical Sciences, New Delhi, India

**Abstract**

Paragangiomas, or glomus tumors, are neoplasms arising from extra-adrenal chromaffin tissue. They frequently cause symptoms by over-production of catecholamines with known predilection to multicentricity. We describe the case of a patient with bilateral carotid body tumor who underwent a preoperative $^{68}$Ga labeled $[1,4,7,10$-tetraazaacyclododecane-$1,4,7,10$-tetraacetic acid]-1-Nal$-$^{3}$-Octreotide ($^{68}$Ga-DOTANOC) positron emission tomography/computed tomography (PET/CT) imaging for staging. This is a unique case in which multiple paraganglioma and pheochromocytoma were demonstrated in a single patient using $^{68}$Ga-DOTANOC PET/CT.

© 2013 Elsevier España, S.L.U. and SEMNIM. All rights reserved.

**Keywords:** $^{68}$Ga-DOTANOC, $^{68}$Ga-tetraazaacyclododecane tetraacetic acid-Nal(3)-octreotide, PET/CT, Paraganglioma, Carotid body tumor, Radionuclide imaging

**Introduction**

Carotid body tumors (CBTs) are the most common head and neck paragangliomas. They arise from neural crest and are known to express somatostatin receptors (SSTRs) which can be exploited for the purpose of imaging with SSTR based positron emission tomography/computed tomography (PET/CT) using $^{68}$Ga labeled octreotide analogs. We present one such case where $^{68}$Ga labeled $[1,4,7,10$-tetraazaacyclododecane-$1,4,7,10$-tetraacetic acid]-1-Nal$-$^{3}$-Octreotide ($^{68}$Ga-DOTANOC) PET/CT was helpful in detecting the primary tumors along with multiple other synchronous lesions.

**Case report**

A 46-year-old male presented with bilateral painless and pulsatile neck masses. His physical examination revealed elevated blood pressure (~190/100 mm of Hg). However, there were no episodes of palpitation, sweating or headache. A contrast enhanced computed tomography study (CECT) of neck was done which revealed bilateral enhancing cervical soft tissue tumors in the region of carotid bifurcation and was diagnosed to be bilateral head and neck paragangliomas (carotid body tumors). A carotid angiogram showed splaying of carotid vessels. Surgical removal

* Corresponding author.

E-mail addresses: rkphulia@hotmail.com, rkphulia@yahoo.com (R. Kumar).

**Estudio preoperatorio con receptores de la somatostatina $^{68}$Ga-DOTANOC PET/TC que demuestra múltiples lesiones sincrónicas en un paciente con paraganglioma de cabeza y cuello**

**Resumen**

Los paragangliomas o tumores de glomus son tumores derivados del tejido cromafín extraadrenal y con frecuencia causan síntomas por exceso de producción de catecolaminas con conocida predilección a ser multicéntricas. Se describe a un paciente con un tumor del cuerpo carotídeo bilateral al que se realizó un estudio PET/TC de extensión preoperatorio con $^{68}$Ga-DOTANOC. Este es un caso único en el que el mismo paciente se le diagnosticó un paraganglioma múltiple y un feocromocitoma con $^{68}$Ga-DOTANOC PET/TC.

© 2013 Elsevier España, S.L.U. y SEMNIM. Todos los derechos reservados.
Verification for the presence of glomus jugulare tumor was done on magnetic resonance (MR) imaging. Thus the localized nature of the head and neck paraganglioma was transformed into a generalized multifocal disease. Also histopathologic assessment of these lesions was not feasible as the patient refused biopsy because of risk of bleeding. The patient was put on a combination of three antihypertensives (propranolol, phenoxybenzamine and amlopidine) and the blood pressure was controlled. Based on the findings of $^{68}$Ga-DOTANOC PET/CT the patient was advised for peptide receptor radionuclide therapy (PRRT) with $^{177}$Lu-DOTATATE. But PRRT was not available in our country at that moment and the patient was unable to afford treatment at any foreign center. He was therefore put on long acting somatostatin analog (sandostatin LAR) along with antihypertensives and he showed stable blood pressure. He was lost to follow up after 8 months.

**Discussion**

Carotid body tumors are the most common head and neck paragangliomas. Owing to their parasympathetic origin they remain mostly asymptomatic and usually produce local symptoms.\(^1\) These tumors are known to be multifocal in certain conditions, especially in familial forms of the disease, and detection of the entire tumor burden is essential from a treatment point of view.\(^2\) $^{111}$In-DTPA-octreotide (Octreoscan), a single-photon emission computed tomography (SPECT) agent for somatostatin receptor scintigraphy (SRS), was considered the gold standard functional imaging for imaging paragangliomas, with an imaging sensitivity of 94%.\(^3\) However, it is not without potential drawbacks. Smaller lesions can be missed because of the limited SPECT resolution; the high physiological uptake in the liver and spleen may mask their involvement and results are generally available only after 48 h. Radiolabeled meta-iodo-benzylguanidine, MIBG (I-123/I-131) is another useful functional imaging modality in the localization of these tumors. Incidental detection of multifocal paragangliomas with radiolabeled MIBG has been described in the past.\(^4\) However, its sensitivity and specificity are quite low since MIBG concentrates only in functioning paragangliomas.

Role of SSTR PET/CT using the tracer $^{68}$Ga labeled octreotide analogs has been described in the past in evaluation of patients with neuroendocrine tumors with a high diagnostic accuracy.\(^5\) It has also shown a very high sensitivity and specificity in the evaluation of patients with pheochromocytomas and paragangliomas with a significant impact on patient management,\(^6\) especially in patients with head and neck paragangliomas. In a previous study on five patients with CBTs, there was detection of additional lesion in three patients.\(^7\) Although detection of occult metastases and presence of additional paragangliomas have been described previously in patients with head and neck paragangliomas using this tracer, the concurrent association of adrenal pheochromocytomas with head and neck paragangliomas has not been documented so far using $^{68}$Ga-DOTANOC PET/CT. If undetected, these occult lesions would continue to serve as source of tumor recurrence and patient morbidity. This case therefore further reiterates the importance of pre-operative work-up of patients with head and neck paragangliomas using $^{68}$Ga-DOTANOC PET/CT. Another advantage of imaging such patients with $^{68}$Ga-DOTANOC PET/CT would be the application of peptide radioreceptor therapy (PRRT) in cases where lesions show uptake on PET imaging and are either surgically unresectable or metastatic in nature.

![Fig 1. $^{68}$Ga-DOTANOC PET whole body maximum intensity projection (MIP) image. Multiple areas of abnormally increased radiotracer uptake are seen throughout the body in different locations marked from 1 to 8. Highest uptake of the tracer was noticed in the neck region corresponding to the carotid body tumors. Physiologic distribution of the tracer in the liver, spleen, kidneys and urinary bladder can be well appreciated.](image-url)
Fig. 2. Axial PET/CT images of the pathologic lesions are shown. Intense tracer uptake is noted in bilateral carotid body tumors [A, arrows]. A small area of focal tracer uptake is also seen in the left jugular canal suggestive of a glomus jugulare tumor [B, bold arrow]. Two areas of abnormal tracer uptake are seen in the thorax in the middle mediastinum [C, bold arrow] and posterior to the left atrium [D, arrow]. Abnormal uptake is also noted in the right peri-hepatic space [E, arrow]. Bilateral adrenal gland neoplasms were also discovered with the right adrenal showing a slight nodularity [F, arrow] while a distinct tumor is seen in the left adrenal [G, arrow]. A total of eight abnormal lesions are described in these PET/CT sections (1–8) corresponding to those appreciated on the MIP image.
Fig. 3. Contrast enhanced CT (CECT) exam of the neck reveals bilateral large soft tissue density masses in the region of carotid space (A, bold arrows). Axial sections through the abdomen are shown (B and C) displaying the adrenal lesions. The left adrenal gland appears rounded with a central hypodense focus (B, bold arrow). No definite abnormality can be appreciated in the right adrenal, except for slight nodularity of the medial limb (C, bold arrow).

References