



⁶⁸Ga-DOTATATE Uptake in Rare Metastatic/Inoperable Mediastinal Paraganglioma with Anomalous Coronary Supply

Anormal Koroner Beslenmeye Sahip Nadir Metastatik/İnoperabl Mediastinal Paragangliomada ⁶⁸Ga-DOTATATE Tutulumu

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Abstract

Mediastinal paragangliomas (PGLs) are extremely rare neuroendocrine tumors. Clinical presentation varies and tumor resection can be challenging due to bleeding and risk of catecholamine surges in functional tumors. We report on a case 68 with a DOTATATE avid mediastinal mass which was histopathologically confirmed as a PGL that was fed by the coronary artery in whole-body positron emission tomography/computed tomography imaging using gallium-68-DOTATATE.

Keywords: Paraganglioma, neuroendocrine tumor, ⁶⁸Ga-DOTATATE, ¹⁷⁷Lu-DOTATATE, PET/CT

Öz

Mediastinal paragangliomalar (PGLs) son derece nadir görülen nöroendokrin tümörlerdir. Klinik tablo değişkenlik gösterir ve fonksiyonel tümörlerde kanama ve katekolamin artışı riski nedeniyle tümör rezeksiyonu zor olabilir. Bu yazıda, galyum-68-DOTATATE kullanılarak yapılan tüm vücut pozitron emisyon tomografisi/bilgisayarlı tomografi görüntülemesinde koroner arter tarafından beslenen bir PGL olarak histopatolojik olarak doğrulanan, DOTATATE tutulumu gösteren bir mediastinal kitle olgusunu sunuyoruz.

Anahtar Kelimeler: Paraganglioma, nöroendokrin tümör, ⁶⁸Ga-DOTATATE, ¹⁷⁷Lu-DOTATATE, PET/BT

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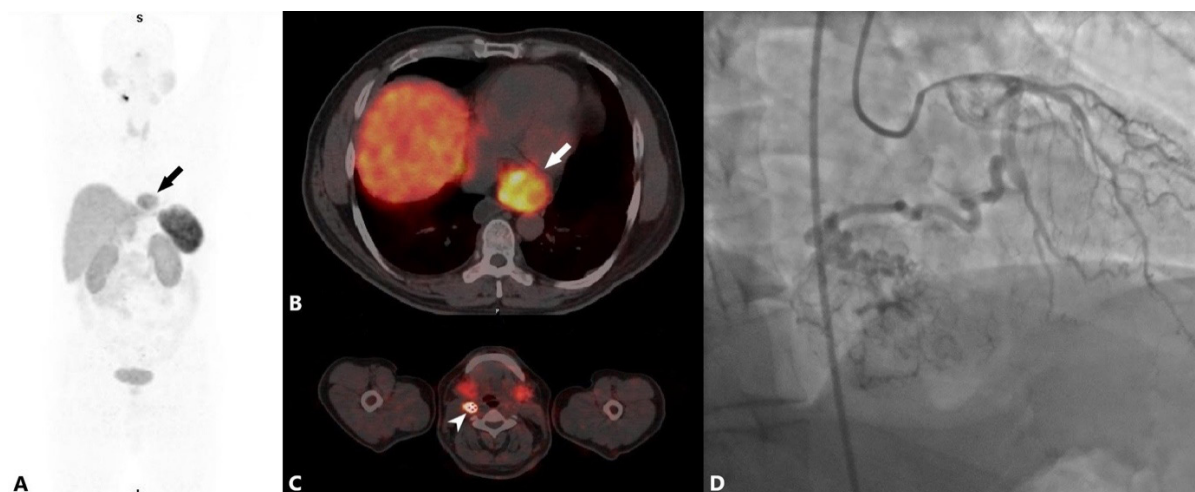


Figure 1. A 52-year-old male patient with significantly elevated normetanephrine level presenting with headache and hypertension was referred for gallium-68 DOTATATE positron emission tomography/computed tomography (PET/CT) for further evaluation. The maximum intensity projection (A) and axial PET/CT (B and C) images revealed a high DOTATATE uptake in the mediastinal mass (black and white arrows; maximum [standard uptake values (SUV_{max}): 18.34] and lymph node at right level 2 (arrowhead; SUV_{max} : 56.56). A thoracotomy was performed to remove the mass, which was determined to be a paraganglioma by histopathology, but it was deemed inoperable because of proximity to major thoracic structures and fed by the coronary artery (D).

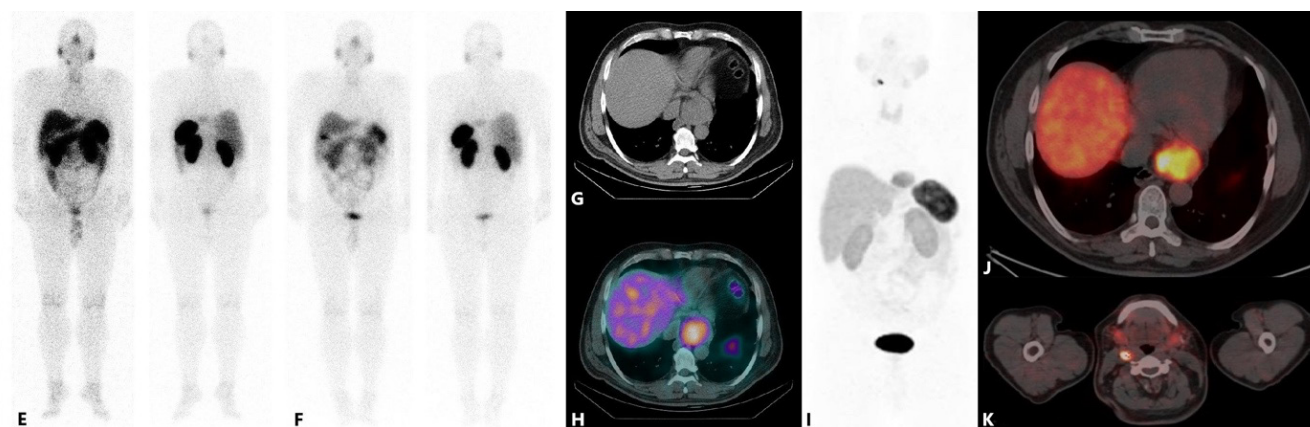


Figure 2. Following embolization of the feeder branch using particles and four cycles of peptide receptor radionuclide therapy (PRRT) with lutetium-177 DOTATATE (^{177}Lu -DOTATATE) [whole-body scans after PRRT, (E) for first cycle and (F) for fourth cycle]. After 4 cycles of ^{177}Lu -DOTATATE, computed tomography (CT) (G) and single photon emission tomography/CT axial fusion (H) images of mediastinal mass. After treatment uptake of cervical lesion was decrease while mediastinal mass uptake was stable. Post-PRRT gallium-68 DOTATATE positron emission tomography/CT [^{68}Ga -DOTATATE PET/CT] maximum intensity projection (I) and axial fusion (J and K) images of mediastinal mass and cervical lymph node. ^{68}Ga -DOTATATE PET/CT imaging in March 2025, 9 months following to fourth cycle, showed partial decreased in tracer avidity with in cervical lymph node [maximum standard uptake values (SUV_{max}): decreased from 56.56 to 33.19; Krenning score of 4 to a score of 3]. Mediastinal mass was stable (SUV_{max} minimal decreased from 18.34 to 17.49; Krenning score of 3 to a score of 3). According to PERCIST criteria, the patient classified stable disease. The serum norepinephrine decreased from 432 to 282 nmol/L after 4 cycles. The patient's symptoms were improved during follow-up. Mediastinal paragangliomas (PGLs) are extremely rare, accounting for approximately 1% to 2% of all PGLs. In patients with inoperable or metastatic PGL, symptomatic or progressive disease is usually treated with chemotherapy, radionuclide therapy, external radiotherapy or tyrosine kinase inhibitors. Functional mediastinal PGLs with anomalous coronary considered high-risk surgically. ^{177}Lu -DOTATATE therapy is safe modality for inoperable and metastatic mediastinal PGLs. ^{68}Ga -DOTA-coupled peptides can be used for imaging because PGLs, which are neuroendocrine tumors arising from pluripotent neural crest stem cells, express somatostatin receptors. Since neuroendocrine tumors overexpress somatostatin receptor like PGLs, ^{68}Ga -DOTATATE PET/CT has a high specificity in detecting them. With the use of DOTATATE's theranostic qualities, patients who may be suitable for PRRT can be chosen using ^{68}Ga -DOTATATE PET/CT (1-7). This report highlights the role of DOTA-coupled peptides in the diagnostic and therapeutic applications of a rare patient with a mediastinal PGL with an unusual coronary artery supply, which poses a high surgical risk.

Ethics

Informed Consent: The written informed consent was obtained from the patient.

Authorship Contributions

Concept: B.O., Design: B.O., Data Collection or Processing: R.Y., M.Ö., Analysis or Interpretation: R.Y., M.Ö., Literature Search: B.O., R.Y., Writing: B.O.

Conflict of Interest: The authors declare no conflicts of interest.

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