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Rapidly growing chest wall mass in a case of sporadic metastatic paraganglioma: imaging with four different PET radiopharmaceuticals

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Abstract

Pheochromocytomas/paragangliomas are rare tumors and mostly benign. We report on a 32 year-old female patient with metastatic paraganglioma who was first diagnosed with an abdominal paraganglioma at the age of 12. She soon developed metastatic disease and received several treatments including external beam radiation and chemotherapy. When she was referred to our institution in 2014 her major complaint was a rapidly growing chest-wall mass on the left side. The patient was imaged at our institution with 4 different positron emission tomography (PET) radiopharmaceuticals.

Keywords

pheochromocytoma; paraganglioma; metastatic; positron emission tomography

Thirty-two year-old female patient with known metastatic paraganglioma (PGL). The patient was referred to our center for further evaluation with a rapidly growing chest mass on the left side. Positron emission tomography (PET) scans with 4 different radiopharmaceuticals

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were performed as well as contrast enhanced computed tomography (CT). The CT images (E) showed a very large, well vascularized soft tissue mass (arrows) following the space between the left 7th and 8th ribs, measuring $19 \times 7.5 \times 5.5$ cm. ¹⁸F-fluoro-2-deoxy-D-glucose (¹⁸F-FDG) (B), which is unspecific [1], but currently the radiopharmaceutical of choice for metastatic PGL [2] due to its far superiority over ¹²³I-metaiodobenzylguaninine (MIBG) [3], and supposed to be an indicator for tumor dedifferentiation [4], showed an intense uptake in this lesion as well as in scattered bone lesions, multiple retroperitoneal lesions, and a large pelvic mass. ¹⁸F-L-dihydroxyphenylalanine (¹⁸F-FDOPA) which is a specific and also very established radiopharmaceutical in functional imaging of PGLs with a known good detection rate especially for sporadic metastatic compared to hereditary PGL [5], showed the same distribution (C) as well as ⁶⁸Ga-DOTATATE (A). This is a new investigational radiopharmaceutical in PGL imaging addressing predominantly somatostatin receptor 2 [6], which is known to be overexpressed in most PGLs [7]. ⁶⁸Ga-DOTATATE PET/CT has recently shown very impressive results in imaging of sympathetic and parasympathetic PGLs [8–11], especially in patients with metastatic disease and underlying succinate dehydrogenase subunit B (*SDHB*) mutation [12] as well as in patients with parasympathetic head and neck PGLs [13, 14]. Positivity on ⁶⁸Ga-DOTATATE PET provides also useful information about the possibility for a future treatment with ¹⁷⁷Lu-DOTATATE. ¹⁸F-fluorodopamine (¹⁸F-FDA), also a very specific and established radiopharmaceutical in PGL imaging addressing the catecholamine synthesis and storage pathways [15] was almost negative for all lesions (D). The identical imaging signature of the chest wall mass and the other metastatic lesions in all 4 different functional imaging modalities made the chest wall mass very suggestive for being a PGL metastases. Since the patient became more and more symptomatic, she finally underwent surgical resection of the chest-wall mass, which was histologically consistent with metastatic PGL and confirmed the imaging findings.

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FIGURE 1.