Recurrent malignant pheochromocytoma with unusual omental metastasis: ^{68}Ga-DOTANOC PET/CT and ^{131}I-MIBG SPECT/CT scintigraphy findings

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ABSTRACT

Pheochromocytomas are rare catecholamine-secreting tumors derived from the sympathetic nervous system. The most common sites of metastasis for pheochromocytoma or extra-adrenal paraganglioma are lymph nodes, bones, lungs, and liver. Patients with known or suspected malignancy should undergo staging with computed tomography (CT) or magnetic resonance imaging as well as functional imaging (e.g. with ^{123}I/^{131}I-MIBG (^{131}I-metaiodobenzylguanidine) and ^{68}Ga-DOTANOC (^{68}Ga-labeled [1,4,7,10-tetraazacyclododecane-1,4,7,10-tetraacetic acid]-1-NaI3-octreotide) positron emission tomography (PET)/CT) to determine the extent and location of disease. We present a case of recurrent malignant pheochromocytoma with unusual site of metastasis in omentum, which was positive on ^{68}Ga-DOTANOC PET/CT and ^{131}I-MIBG single-photon emission computed tomography (SPECT/)/CT scintigraphy.

Keywords: ^{131}I-MIBG, ^{68}Ga-labeled [1,4,7,10-tetraazacyclododecane-1,4,7,10-tetraacetic acid]-1-NaI3-octreotide, malignant pheochromocytoma, positron emission tomography/computed tomography, Single-photon emission computed tomography/computed tomography

A 49-year-old male who had undergone right adrenalectomy for pheochromocytoma 8-years ago presented with impaired glucose tolerance with diabetes and hypertension since 1 year. His 24 h urine vanilylmandelic acid was elevated in range of 233 mg/24 h (normal: 0-13.6 mg/24 h). His computed tomography (CT) abdomen revealed multiple intra-abdominal masses in right suprarenal, perirenal, retrocaval, peripancreatic, and in omentum with vivid arterial enhancement. The patient was referred for ^{68}Ga-labeled [1,4,7,10-tetraazacyclododecane-1,4,7,10-tetraacetic acid]-1-NaI3-octreotide (DOTANOC) positron emission tomography (PET)/CT study for restaging purpose. ^{68}Ga-DOTANOC PET/CT revealed large aortocaval lymph nodal mass (6.4 × 5.3 cm) with area of necrosis and DOTANOC uptake in non-necrotic part (SUV max-3.1). Multiple peripancreatic, mesenteric, and retroperitoneal lymph nodes were noted with increased tracer uptake. It also showed multiple omental deposit with increased tracer uptake (SUVmax-2.5) [Figure 1]. ^{131}I-metaiodobenzylguanidine (MIBG) planer whole body scintigraphy was performed to evaluate the therapeutic potential of ^{131}I-MIBG in view of the metastatic nature and inoperability of the disease. ^{131}I-MIBG study showed increased tracer uptake in upper and mid abdomen regions. Single-photon emission computed tomography (SPECT)/CT study revealed MIBG concentrating lesions involving aortocaval, multiple peripancreatic, mesenteric and retroperitoneal lymph node, and omental deposits suggestive of recurrent disease [Figure 2]. Ultrasound-guided aspirate from right suprarenal lesion and omental lesion showed cytomorphological feature compatible with pheochromocytoma [Figure 3]. As the lesions were showing more ^{131}I-MIBG uptake than ^{68}Ga-DOTANOC uptake, the patient was taken for ^{131}I-MIBG therapy.

Pheochromocytomas are rare catecholamine-secreting tumors derived from chromaffin cells. In all, 10-50% of intra-abdominal extra-adrenal paraganglioma are malignant.[1] Metastatic spread is the only reliable criterion for the diagnosis of malignant pheochromocytoma. In 7% of the cases, metastasis occurred in more than one organ.[2,3] The most common sites of metastasis are lungs, liver, lymph nodes, and bones. Previous studies with ^{111}In-Octreotide have shown higher sensitivity for detecting metastatic pheochromocytoma than for detecting benign pheochromocytoma.[4] ^{68}Ga-DOTANOC PET/CT showed high sensitivity for both phaeochromocytoma and paragangliomas. This is partly because of wide spectrum of affinity of ^{68}Ga-DOTANOC for SSTR subtypes. On the other

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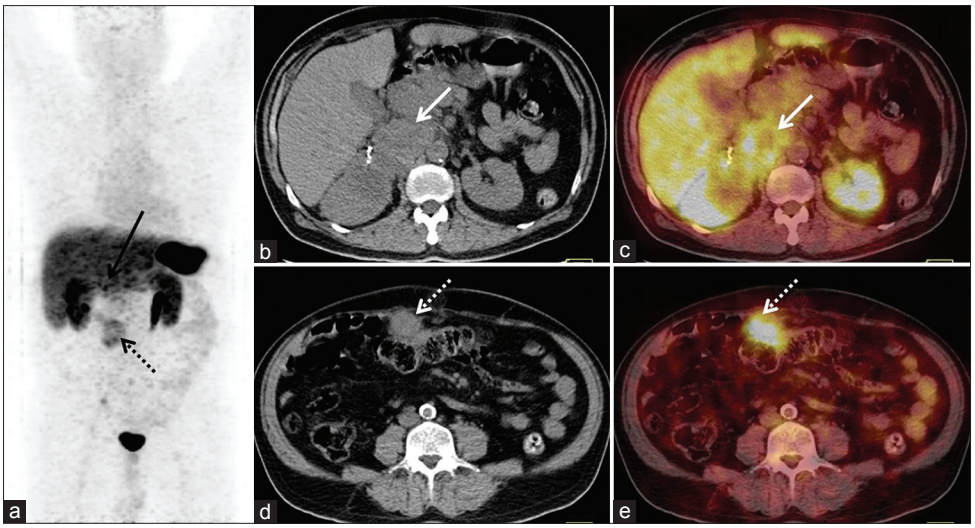

Figure 1: ^{68}Ga-DOTANOC PET/CT revealing large aortocaval lymph nodal mass (6.4 × 5.3 cm) with area of necrosis and DOTANOC uptake in non-necrotic part (a-c, arrows). It also showed an omental deposit with increased tracer uptake (a, d, e, dotted arrows). ^{68}Ga-DOTANOC = ^{68}Ga-labeled [1,4,7,10-tetraazacyclododecane-1,4,7,10-tetraacetic acid]-1-NaI3-octreotide, PET = Positron emission tomography, CT = Computed tomography

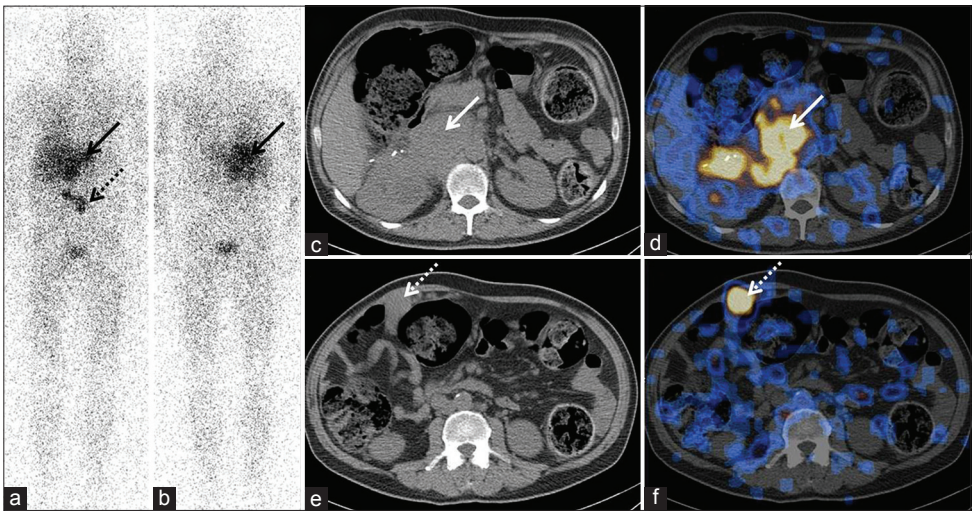

Figure 2: ^{131}I-MIBG planer whole body scintigraphy anterior (a) and posterior (b) images showing increased tracer uptake in upper and mid abdomen regions. SPECT/CT study revealed MIBG concentrating lesions involving aortocaval lymph node (c and d, arrows) and a omental deposit suggestive of recurrent disease (e and f, dotted arrows). ^{131}I-MIBG = ^{131}I-metaiodobenzylguanidine, SPECT = Single-photon emission computed tomography, CT = Computed tomography

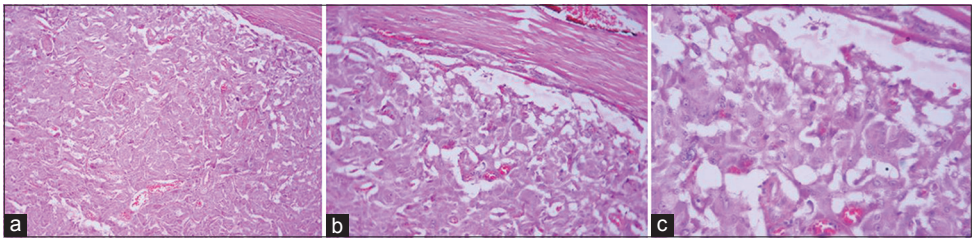

Figure 3: Ultrasound-guided aspirate from right suprarenal lesion and omental lesion (a) (×10), (b) (×20), (c) (×40) shows groups of cell in nesting pattern with abundant eosinophilic cytoplasm and rounded nuclei showing mild nuclear pleomorphism compatible with pheochromocytoma

hand, uptake of ^{131}I-MIBG is dependent on the expression of vesicular monoamine transporters (VMAT 1, 2). Expression of VMAT is high in benign phaeochromocytoma but is reduced in malignant phaeochromocytoma and paragangliomas.[5] In our case study, ^{131}I-MIBG scintigraphy and ^{68}Ga-DOTANOC PET/CT both showed increased tracer uptake in metastatic pheochromocytoma. This case demonstrates unusual site of omental metastasis in malignant pheochromocytoma. Occurrence of the extra-adrenal paragangliomas outside the normal distribution of the paraganglionic tissue can probably be explained by the migratory property of the neural crest cells during embryogenesis.[6,7] These cells can form collection of paraganglionic tissue and give rise to paragangliomas.

The recurrent lesions in our case were positive both on ^{131}I-MIBG scintigraphy and ^{68}Ga-DOTANOC PET/CT. But ^{131}I-MIBG was strongly positive, which altered the patient management, and the patient was taken for ^{131}I MIBG therapy.

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