Case Report

Malignant perivascular epithelioid cell tumor of the ileum on ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography with pathological correlation

ABSTRACT

A 75-year-old woman presented with a 1-month history of abdominal pain. Contrast-enhanced computed tomography (CT) demonstrated a large solid mass in the left lower abdominal quadrant, suspicious for malignancy. Staging with ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/CT imaging demonstrated intense FDG uptake in the mass with no evidence of metastatic disease. Complete surgical resection was performed, and histopathological analysis confirmed a malignant perivascular epithelioid cell tumor of the ileum.

Keywords: ^{18}F-fluorodeoxyglucose, perivascular epithelioid cell tumor, positron emission tomography/computed tomography

INTRODUCTION

Malignant perivascular epithelioid cell tumors (PEComas) are rare mesenchymal tumors that can develop in multiple anatomic sites including the gastrointestinal tract as well as in genitourinary, pulmonary, and musculoskeletal structures. Metastases are present, however, in over one-third of cases of malignant gastrointestinal PEComa, potentially limiting curative treatment options. Therefore, evaluating for the presence of metastatic disease before any definitive resection is critical in the management of these patients. Due to their relative rarity, the imaging features of malignant PEComas are not well described. We describe a rare case of malignant PEComa of the ileum where ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) helped confirm nonmetastatic disease before surgery.

CASE REPORT

A 75-year-old female presented with a 1-month history of abdominal pain and bloating. Initial CT of the abdomen and pelvis with intravenous contrast was performed

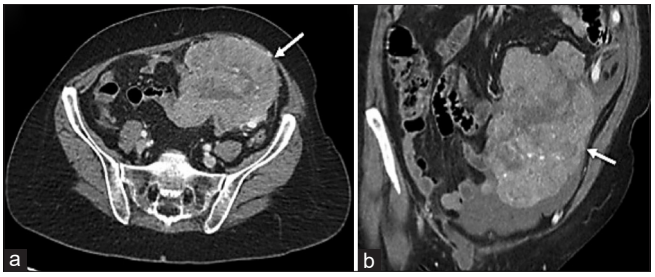

Figure 1: Axial (a) and coronal (b) computed tomography of the abdomen and pelvis with intravenous contrast demonstrating a solid, heterogeneously enhancing mass in the lower abdomen (arrows) suspicious for malignant neoplasm

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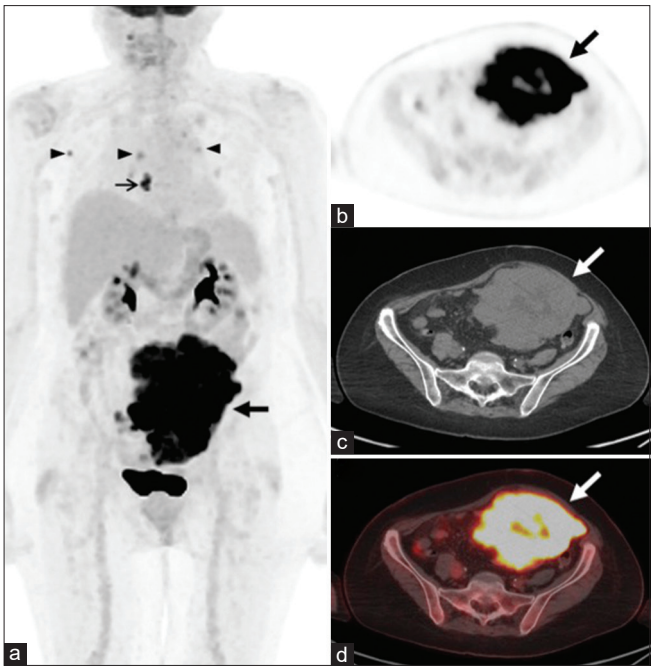

Figure 2: Maximum intensity projection (a), axial positron emission tomography (b), axial nonintravenous contrast-enhanced computed tomography (c), and axial fused positron emission tomography/computed tomography (d) demonstrated an intensely hypermetabolic mass (maximum standardized uptake value: 47) in the left lower quadrant involving loops of small bowel with central foci of necrosis (arrows). Additional foci of uptake in a chest wall inflammatory skin lesion (line arrow) and nonenlarged reactive-appearing thoracic nodes (arrowheads) were noted

showing a mass suspicious for malignancy in the left lower quadrant [Figure 1]. Staging was undertaken with ^{18}F-FDG PET/CT [Figure 2]. There was no evidence of metastatic disease. Initial diagnostic considerations included gastrointestinal stromal tumor (GIST), desmoid tumor, small bowel adenocarcinoma, and lymphoma, and image-guided core-needle biopsy was performed revealing a poorly differentiated malignancy with epithelioid features. Midline laparotomy, complete resection of the pelvic mass with segmental small bowel resection, and entero-enteric anastomosis were subsequently performed. The morphology and immunoprofile of the tumor were consistent with a diagnosis of malignant perivascular epithelioid cell tumor of the ileum [Figure 3].

DISCUSSION

PEComas are a rare histologic family of neoplasms that demonstrate distinctive epithelioid cell histopathologic features and develop in a perivascular distribution, expressing both human melanocytic (HMB-45 and/or melanin A) and smooth muscle markers such as desmin.[1,2] Gastrointestinal tract PEComas are uncommon with PEComas of the small bowel considered exceedingly rare.[3-5] Malignant PEComas must have two of the following criteria: size >5 cm,

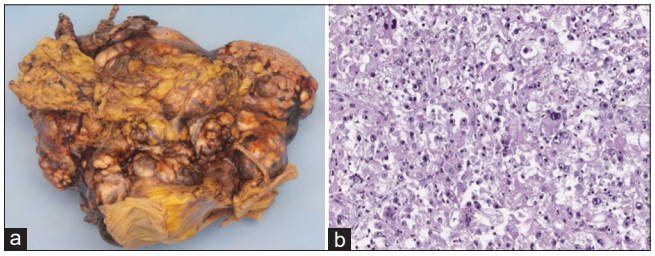

Figure 3: Gross pathological specimen showed a mass involving the ileum (a) and H and E-stained microscopic examination of sampled tissue (b) revealed a high-grade carcinoma composed of large sheets of pleiomorphic, polygonal epithelioid cells with enormous cherry-red nucleoli and clear to granular eosinophilic cytoplasm. Immunohistochemistry was positive for melanin A and cathepsin-K, with patchy HMB45 and focal desmin reactivity. Fumarate hydratase showed retained staining, and TFE3 was weakly positive

mitosis >1/50 high-power fields, high nuclear grade and cellularity, and necrosis or vascular invasion.[2] On imaging, malignant PEComas may mimic other mesenchymal tumors such as GIST and desmoid tumors.[4] On CT, malignant PEComas are usually well-circumscribed, hypo or iso-attenuating to muscle with necrotic components.[6] Whereas benign PEComas usually demonstrate minimal or no FDG uptake, malignant PEComas[7,8] are often intensely FDG avid, possibly related to upregulation of mammalian target of rapamycin pathway which controls glucose transporter 1 function.[9] Although surgery is the main treatment option for the management of localized malignant gastrointestinal PEComas, metastases are seen in up to 37% of cases[3] potentially altering management with patients receiving systemic therapies instead.[3]

CONCLUSION

To the best of our knowledge, we present the first case of malignant PEComa of the ileum on ^{18}F-FDG PET/CT and describe the clinical utility of PET/CT in initial staging of primary malignant small bowel PEComa before operative management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Hornick JL, Fletcher CD. PEComa: What do we know so far? Histopathology 2006;48:75-82.
2. Folpe AL, Mentzel T, Lehr HA, Fisher C, Balzer BL, Weiss SW. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: A clinicopathologic study of 26 cases and review of the literature. Am J Surg Pathol 2005;29:1558-75.
3. Doyle LA, Hornick JL, Fletcher CD. PEComa of the gastrointestinal tract: Clinicopathologic study of 35 cases with evaluation of prognostic parameters. Am J Surg Pathol 2013;37:1769-82.
4. Iwamoto R, Kataoka TR, Furuhata A, Ono K, Hirota S, Kawada K, *et a*l. Perivascular epithelioid cell tumor of the descending colon mimicking a gastrointestinal stromal tumor: a case report. World J Surg Oncol. 2016;14;1:285.
5. Lu B, Wang C, Zhang J, Kuiper RP, Song M, Zhang X, *et al*. Perivascular epithelioid cell tumor of gastrointestinal tract: Case report and review of the literature. Medicine (Baltimore) 2015;94:e393.
6. Phillips CH, Keraliya AR, Shinagare AB, Ramaiya NH, Tirumani SH. Update on the imaging of malignant perivascular epithelioid cell tumors (PEComas). Abdom Radiol (NY) 2016;41:368-76.
7. Ciarallo A, Makis W, Hickeson M, Derbekyan V. Malignant perivascular epithelioid cell tumor (PEComa) of the uterus: Serial imaging with F-18 FDG PET/CT for surveillance of recurrence and evaluation of response to therapy. Clin Nucl Med 2011;36:e16-9.
8. Wu J, Jiang L, Zhang F, Huang Y, Wang H. Malignant perivascular epithelioid cell tumor of lung on FDG PET/CT. Clin Nucl Med 2019;44:469-71.
9. Sun L, Sun X, Li Y, Xing L. The role of (18)F-FDG PET/CT imaging in patient with malignant PEComa treated with mTOR inhibitor. Onco Targets Ther. 2015;30:1967-70.