Case Report

Malignant perivascular epithelioid cell tumor of the ileum on 18F-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 18F-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: 18F-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 18F-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

Jeeban Paul Das, Jad Bou-Ayache, Marc J. Gollub, Christopher C. Riedl, Gary A. Ulaner
Department of Radiology, Memorial Sloan Kettering Cancer Centre, New York, USA
Address for correspondence: Dr. Jeeban Paul Das, Memorial Sloan Kettering Cancer Center, 1275 York Ave, Box 77, New York, NY 10065, USA.
E-mail: dasj@mskcc.org
Submission: 16-Aug-2020, Revised: 28-Sep-2020, Accepted: 09-Sep-2020, Published: 23-Oct-2020

How to cite this article: Das JP, Bou-Ayache J, Gollub MJ, Riedl CC, Ulaner GA. Malignant perivascular epithelioid cell tumor of the ileum on 18F-fluorodeoxyglucose positron emission tomography/computed tomography with pathological correlation. World J Nucl Med 2021;20:208-10.
Das, et al.: PEComa of the ileum

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

CONCLUSION

To the best of our knowledge, we present the first case of malignant PEComa of the ileum on ¹⁸F-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.

Financial support and sponsorship

This research was supported in part by the NIH/NCI Cancer Center Support Grant P30 CA009748.

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 al. 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, Ramaia NY, 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.