

Uterine Endometrial Stromal Sarcoma on MRI and ¹⁸F-FDG PET/CT: A Case Report with Literature Review

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Abstract

A rare case of endometrial stromal sarcoma is presented with the features of MRI and ¹⁸F-FDG PET/CT, as well as a brief discussion with a literature review. The findings of our case and the few other reports open the possibility for evaluating the usefulness of the PET/CT findings as prognostic factors in these patients as it might predict tumor biology.

Keywords

Endometrial stromal sarcoma, ¹⁸F-FDG PET/CT, Tumor biology

Introduction

Endometrial stromal sarcomas (ESS) are a rare group of malignant mesenchymal tumors arising from the connective tissue of the endometrium, which resemble cells in their proliferative stage. These tumors represent 6–20% of uterine sarcomas and less than 1% of primary uterine malignancies; being low-grade, the most frequently diagnosed. Diagnosis is based on surgical pathology, but imaging plays a very important role in staging, considering that approximately 30–50% of patients have extra uterine spread at the time of diagnosis, and this determines treatment. MRI can determine with high accuracy the extension and invasion of the lesion, but ¹⁸F-FDG PET/CT is more accurate for the detection of abnormal lymph nodes and distant metastases, thus combining these two modalities is the best approach.

¹⁸F-FDG PET/CT findings also might be considered as prognostic factors as they can predict tumor biology; however, there are few case reports of ESS and its behavior in ¹⁸F-FDG PET/CT. We present a case of a 67 years old woman in whom, although the initial biopsy reported a probable low-grade lesion, PET/CT findings suggested a more aggressive tumor, which was finally confirmed. Our findings and few other reports open the possibility for evaluating deeper the usefulness of PET/CT in these patients.

Case Report

A 67-year-old woman, with no relevant history, presents with low abdominal pain and vaginal bleeding. She was first evaluated in another institution where transvaginal ultrasound reported a suspicious endometrial mass. Ca-125 was in the normal range (21 U/ml) and endometrial aspiration cytology was reported negative for malignancy. She was referred to our institution where MRI was performed, finding a 11.8 cm sized well defined cystic and solid uterine mass involving the endometrium and myometrium, with heterogeneous T2-WI signal intensity and heterogeneous contrast enhancement and diffusion restriction in the solid component. Multiple retroperitoneal enlarged lymph nodes were also identified and a small amount of pelvic ascites (Figure 1).

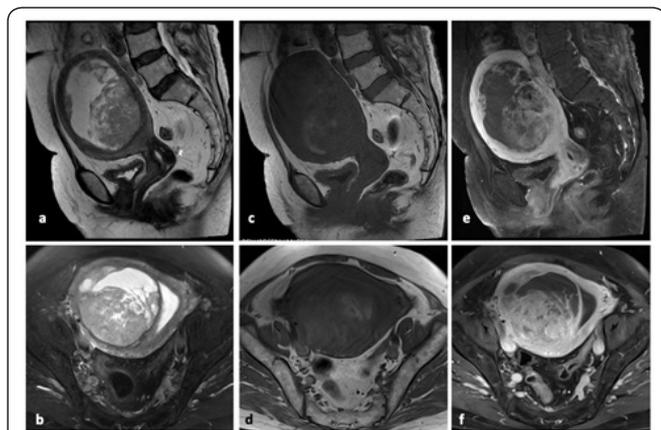


Figure 1: MRI a-b) sagittal and axial T2-WI, c-d) sagittal and axial T1-WI and e-f) sagittal and axial T1+C, showing a 11.8 cm sized well defined complex uterine mass with cystic and solid components, involving the endometrium and myometrium, with heterogeneous signal intensity and heterogeneous contrast enhancement within the solid component.

Endometrial biopsy was reported as suggestive of low-grade sarcoma. ¹⁸F-FDG PET/CT was requested for pre-surgical staging and findings included markedly increased FDG uptake in the solid component of the heterogeneous malignant mass, with SUV_{max} value of 14.4, and multiple enlarged hypermetabolic metastatic retroperitoneal lymph nodes in bilateral iliac, aortocaval, periaortic and retropancreatic areas, with SUV_{max} value of 20; with this findings we suggested the possibility of a higher grade lesion (Figure 2).

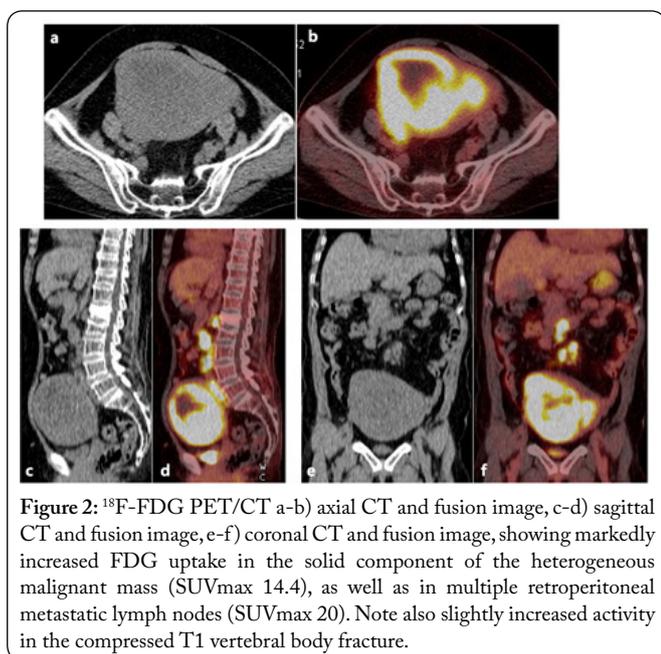


Figure 2: ¹⁸F-FDG PET/CT a-b) axial CT and fusion image, c-d) sagittal CT and fusion image, e-f) coronal CT and fusion image, showing markedly increased FDG uptake in the solid component of the heterogeneous malignant mass (SUV_{max} 14.4), as well as in multiple retroperitoneal metastatic lymph nodes (SUV_{max} 20). Note also slightly increased activity in the compressed T1 vertebral body fracture.

The patient was taken to surgery and the final pathology report confirmed a malignant spindle cell neoplasm consistent with high-grade uterine stromal sarcoma, CD10 positive, smooth muscle actin (SMA) positive, estrogen and progesterone receptors negative, with ki67 positive in 60%.

Discussion

Endometrial stromal sarcomas (ESS) are a rare group of

malignant mesenchymal tumors arising from the connective tissue of the endometrium, which resemble cells in their proliferative stage. They were first described in 1966 by Norris and Taylor and since then they have been classified into low grade ESS, high grade ESS and undifferentiated ESS, based on the presence or not of normal proliferative endometrial stroma [1]. The latest WHO classification included endometrial stromal nodule (ESN), which represents disease with no or minimal myometrial invasion (< 3mm) and no lymphovascular invasion [2].

The most common location is the uterus, but they can also affect the cervix, and other rare described locations include the ovaries, fallopian tube, pelvic cavity or peritoneum and colon, all related to endometriosis [3]. They typically affect postmenopausal women, with a mean age at diagnosis between 42 and 58 years. Only 10 - 25% of affected women are premenopausal [4, 5].

They can be related, as other gynecological malignancies, to hyperestrogenism and some cases have been reported in patients with polycystic ovary syndrome [6]; tamoxifen therapy and radiation for other malignancies have been linked too [7].

Multiple genetic abnormalities have been detected in women with this type of sarcomas. The most associated to low-grade tumors are related to JAZF11-SUZ12 gene and for high grade and undifferentiated sarcomas, abnormalities have been detected in BCOR ZC3H7B-BCOR and YWHAE-FAM22 respectively [8, 9].

Clinical presentation includes low abdominal pain and vaginal bleeding, but up to 25% of cases can be asymptomatic [10].

Diagnosis is based on surgical pathology because curettage specimens have shown low diagnostic accuracy. Also, because morphologic, immunohistochemical and molecular features can be very similar and growth pattern and lymphovascular invasion must be determined [2]. These tumors tend to have estrogen and progesterone receptors, and even there are no specific tumor markers, CD10 can be helpful to distinguish them from leiomyomas as well as asdesmin and h-caldesmon receptors [11, 12].

Morphological imaging findings include irregular margined intra-myometrial nodule or mass, usually large in size or with multiple myometrial nodular or wormlike extensions. Echogenicity, density and signal intensity are non-specific and usually heterogeneous due to internal necrosis, hemorrhage and areas of preserved myometrium inside the mass. MR has proven to be the most useful tool in the pretreatment survey of uterine malignancies overall because it can determine with high accuracy the extension and invasion of the lesion. On MR images, the lesion's solid components usually show slight hypointensity on T1-weighted images and slight hyperintensity with mottled isointensity on T2-weighted images. Enhancement is heterogeneous but prominent because of high vascularity, as well as diffusion restriction due to high cellularity. Some of these features are useful to differentiate leiomyomas with different grades of degeneration from ESS [13]. Imaging differential diagnosis

include mainly leiomyomas, leiomyosarcomas, endometrial cancer and adenomyosis.

¹⁸F-FDG PET/CT, as a functional hybrid imaging modality, quantifies activity of the tumor and it is being increasingly used for the staging and monitoring of gynecological malignancies in general. However, there are yet few reported cases of ESS and its behavior in ¹⁸F-FDG PET/CT. Umesaki et al. reported in 2001, five cases of patients with uterine sarcomas in whom ¹⁸F-FDG PET/CT was performed in addition to routine morphological images, and they found that 100% of examinations were positive in contrast to MRI and US were only 80% and 40% were positive respectively [14]. In the few reports from the literature, it has been found that, like other sarcomas, ESS presents a significant uptake of FDG in the solid component, which is probably related to increase glucose transporter I expression, hexokinase II and lactic dehydrogenase, just as in other sarcomas [15]. In our case, although the initial biopsy reported a probable low-grade lesion, the PET/CT findings suggested a higher-grade lesion given its significant FDG uptake and extent with multiple metastatic lymph nodes.

Currently, the main role of PET/CT in this patient is for initial staging; where the detection of lymph nodes and distant metastases can change treatment and prognosis of the patient considering that approximately 30-50% of patients have extra uterine spread at the time of diagnosis. It is also indicated for the follow-up of high-risk patients or in patients with suspected relapse due to the presence of symptoms or an unexplained elevation of Ca 125, where it has shown better performance than conventional imaging for detection of recurrent or metastatic lesions [16, 17].

Prognosis primarily depends on the stage of the disease at diagnosis, which is determined by the International Federation of Gynecology and Obstetrics (FIGO) staging. Prognostic factors are still not fully determined because multiple studies have shown different results, but older aged, advanced stage with nodal metastasis, atypia and CD10 low expression were the more associated with poor survival. Low-grade tumors have very good prognosis with a 5-year survival between 50-90% depending on stage; undifferentiated tumors present with high stage disease in more than 60% of cases so they have a very low survival rate; and high grade tumors have a variable prognosis between those two, with the majority of patients having early recurrences (< 1 year) [12, 18]. The findings of our case and the few other reports open the possibility for evaluating the usefulness of the PET/CT findings as prognostic factors in these patients as it might predict tumor biology.

Conflict of Interest

The authors declare no conflict of interest.

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