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An unusual enhanced Doppler vascular profile of a rare uterine tumor: PEComa

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Summary

PEComas represent a rare class of mesenchymal tumors, with different primary locations. There are less than 100 cases of uterine PEComas published in English literature until now and information considering imaging features of these PEComas is very limited, focusing on CT and MRI and not as much on ultrasounds (US). The authors present here a case of rapidly growing uterine PEComa, with local invasive potential and recurrence, and the review of literature on US characteristics of PEComas. Harboring a hyperechogeneous heterogeneous aspect with no clear separation from the adjacent uterus on the whole boundary, with an extremely rich central vascular network, with low impedance and a rapidly growing profile, this tumor does not show the classic US appearance of malignant PEComas, which are generally easily confused with leiomyomas. However, even if this pattern did not allow the authors to anticipate the histopathological result, it offered clear clues about its invasiveness potential.

Key words: uterine PEComa; Doppler vascularisation; Uterine tumor.

Introduction

Perivascular epithelioid cell tumors (PEComas) represent a rare class of mesenchymal tumors, described for the first time by Bonetti *et al.* in 1992 [1] and including entities as: angiomyolipoma, clear cell sugar tumor of the lung and extrapulmonary sites, and lymphangioleiomyoma.

Even if the uterine primary location is a common one, accounting for 25% of all PEComas [2], there are no more than 100 case presentations published in the English literature, most of them focusing on the histopathological diagnosis and clinical prognosis.

Information considering the imaging features of PEComas is very limited and even in these articles, focusing on CT and MRI descriptions and not as much on ultrasound, and usually, benign and malignant PEComas are grouped together [3, 4]. However ultrasound is the first line imaging technique and if the US misinterprets the tumor as leiomyoma, the other most expensive imagistic techniques, as CT, MRI, PET-scan, are not required before surgery [5].

The aim of the present article was to present the US characteristics of a uterine PEComa, rapidly growing and with a high potential of local recurrence, and to make a review of literature on US characteristics of PEComas.

Case Report

A 33-year-old patient, gravid-0, para-0, performed in the present clinic, a vaginal ultrasound for a two-year primary infertility. The patient's past medical history was insignificant. She had no history and no clinical signs of tuberous sclerosis.

On this occasion, a hyperechogenic pseudo-encapsulated tumor of four cm diameter localized in the posterior uterine wall, partially distorting the uterine cavity, was described and interpreted as a type II fibroid. The endometrium and the ovaries were normal (Figure 1). A myomectomy was performed, and the pathological exam confirmed the diagnosis of leiomyoma. Three months after surgery, the patient underwent an endovaginal ultrasound control, which revealed a new uterine tumor, localized intramural posterior with extension into the right broad ligament, hyperechogeneous, heterogeneous, without clear space separation from the uterus on its entire boundary, suspected as a rapidly growing recurrent fibroid or a sarcoma. Doppler ultrasound showed a tumor with an extremely rich central vascular network with low impedance (Figures 2 and 3).

In the uterine cavity, a polypoid image of one cm diameter with vascular pedicle was seen. The ovaries were normal at ultrasound. Surgical reintervention was performed consisting in tumor removal with preservation of the uterus, tumor enucleation being relatively difficult. Macroscopically, the tumor measured 55/66 mm diameter, with yellowish-gray appearance, imprecisely delineated, with no areas of necrosis on cut surface. Intraoperative frozen sections examination revealed no signs of malignancy.

Histopathological examination of formalin fixed and paraffin embedded samples revealed a non-encapsulated tumor proliferation consisting in areas of fascicles of spindle cells alternating with areas of epithelioid-like cells (around small vessels). The tumor cells showed round or oval central nuclei, with reduced pleomorphism, eosinophilic or vacuolated pale cytoplasm, and a low mitotic activity (3 mitosis/10 HPF in the most active areas). No tumor necrosis was observed. The boundary between tumor nodule and adjacent myometrium was irregular and focally infiltrative. Immunohistochemistry revealed weak to moderate positivity for melanocytic (HMB-45, Melan-A) markers with focal

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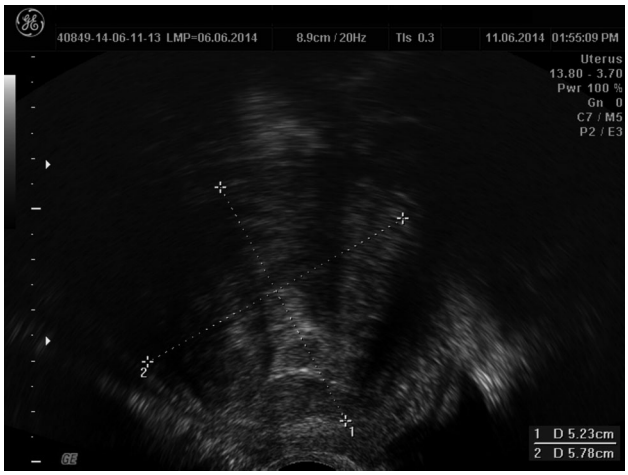

Figure 1. — Image of 2D ultrasound in the uterine PEComa (before first intervention).

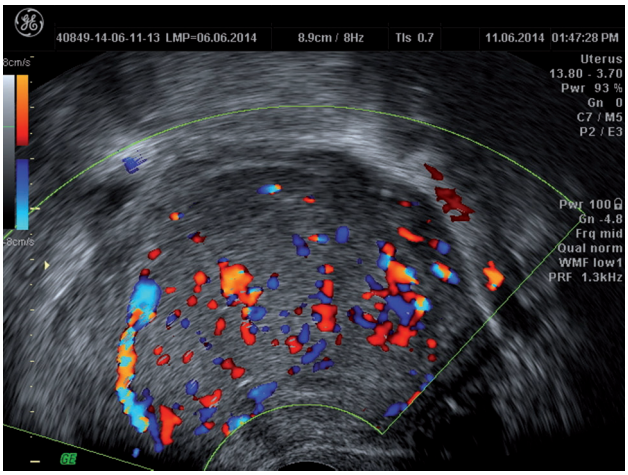

Figure 2. — Doppler aspect of uterine PEComa with local invasive potential and local recurrence.

distribution in tumor cells, diffuse positivity for smooth muscle (SMA, desmin) markers and for estrogen receptor, while Ki67 was positive in 5-10% of tumor cells in the most active areas (Figure 4).

Histopathological features and immunohistochemical profile were consistent with a uterine perivascular epithelioid cell tumor, not otherwise specified (PEComa-NOS).

Considering the size of over four cm of the tumor, the mitotic activity (3 mitosis/10 HPF), the rapid tumor growing rhythm (up to five cm in three months), and the imprecise delineation of the tumor with microscopically confirmed myometrial invasion, the present authors recommended a hysterectomy to the patient, despite her desire for future pregnancy. After a whole body magnetic resonance imaging (MRI) to exclude distant metastasis, the authors performed total hysterectomy with bilateral adnexectomy and pelvic lymphadenectomy.

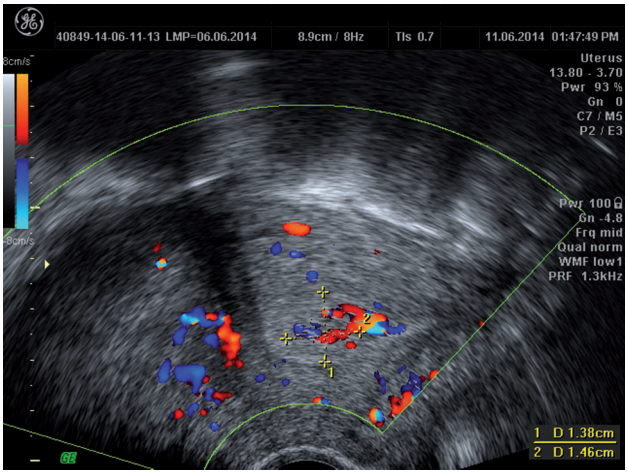

Figure 3. — Endometrial polyp associated to uterine PEComa.

Discussion

Uterine PEComas are tumors with unpredictable prognosis; most of them are benign or show local recurrence, although rare malignant PEComas have been reported either with metastasis after many years of clinical "silence" [6].

Here, the authors have reported a case of uterine PEComa-NOS, rapidly growing, with local invasive potential and local recurrence. They analyzed US characteristics of this tumor, performing a review of literature. There are no more than 11 articles in English literature (discovered at a PubMed search), referring to PEComa ultrasound appearance .

Most US examinations performed before surgery, described PEComas as well circumscribed masses, heteroechoic, with no cystic areas or significant vascularity on Doppler examination, being confused usually with leiomyomas: Tirumani *et al.*, 2014 n= 3/5 cases [3]; Bosincu *et al.* 2005: two cases [7]; Fadare *et al.*, 2004: one case [8]; Liu *et al.*, 2009: one case [9]; and Gan *et al.,* 2007: one case [10].

Some authors described a character of malignancy: Cossu *et al.*, 2004: two cases [2]; Tirumani *et al.*, 2014: 1/5 cases [3]; Manganaro *et al.,* 2002: one case [11]; Issat *et al.*, 2012: one case [12]; and Natella *et al.*, 2014: one case, and stated that these tumors lack specific clinical and radiological features.

In the present case, the rapidly growing pattern, the high vascularity with low impedance, and the imprecise borders of the tumor, raised the suspicion of uterine sarcoma. There was no cystic or hypoechoic area suggesting necrosis. The ultrasound aspect persisted on the second recurrence lesion.

Uterine PEComas have different locations: intramural, on a polypoid adenomyoma, in the uterine cervix, with expansion to the large ligament (as in present case), into the round ligament. They can be associated with other

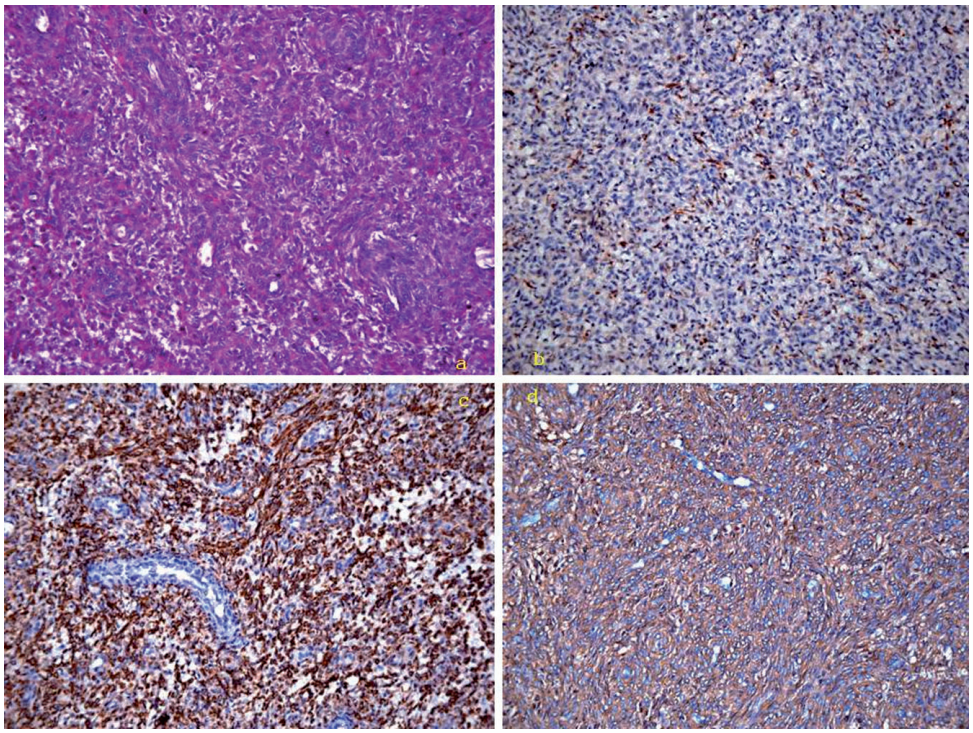

Figure 4. — Immunohistochemical staining in uterine PEComa. a) PEComa-HE staining x200; b) PEComa, HMB45 x200, zonal positive; c) PEComa, desmin x200, diffusely positive; d) PEComa, SMA x200, diffusely positive.

uterine lesions: endometrial polyp, leiomyoma (as in present case).

Conclusion

The ultrasound profile of these tumors does not correlate with the histological type, but this entity must be kept in mind as a differential diagnosis. Instead, ultrasound appearance correlates better with the prognosis of the lesion. The present case with the rapidly growing model, the enhanced Doppler vascularization with low impedance, and the partial infiltrative margins, suggest the beginning local invasiveness and the high recurrence potential of the tumor.

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