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P97. PERIVASCULAR EPITHELIOID CELL TUMOR OF THE UTERUS (PECOMA): A RARE CASE

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Context:
Perivascular epithelioid cell tumor is a rare mesenchymal neoplasm. It originates from the perivascular epithelioid cell line and was first described in 1943 as an abnormal myoblast.
Objective:
These tumors showed an unusual cell type with a perivascular distribution. Moreover, these cells are characterized immuno reactive for melanocytic markers, with an epithelioid appearance and a clear acidophilic cytoplasm, thus the term "perivascular epithelioid" was introduced. The most commonly found in women, which originate in many different body tissues the most common being the GI/GU tracts and the retroperitoneum. The uterus is the most prevalent site of involvement but fewer than 50 uterine PEComas have been reported in the literature.
Method:
We present a case of uterine pecoma with abnormal uterine bleeding.
Patient:
A 44-year-old woman presented to our clinic with pelvic tenderness upon physical examination and abnormal menstrual bleeding.
Intervention:
CT scans showed a mass at the uterine wall sized 4 cm with no other pathology. She had a free medical history with no prior surgeries. The patient underwent a dilation and curettage prior to abdominal myomectomy.
Main Outcome Measure:
The result of endometrial biopsy is secretory endometrium. Histopathology report showed a neoplasm of mesenchymal origin, with immunophenotypical characteristics compliant with Perivascular Epithelioid Cell tumor. The cells showed a mitotic rate < 1/10 HPF. The necrosis and atypic cells were not detected. Immunohistochemistry report was positive for the melanocytic marker HMB-45 and Melan A, vimentin, SMA, CD-10, S-100 positive and caldesmon marker was rare positive.
Result:
In combination of these pathological features and the Immunohistochemistry results, the final pathologic diagnosis was of benign perivascular epithelioid cell tumor (PEComa).

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Conclusions:
In most of the cases, PEComa presents with abnormal vaginal bleeding, abdominal pain and uterine symptoms. Six high-risk criteria have been proposed for the classification of PEComas: tumor size â‰¥ 5 cm, infiltrative growth pattern, high nuclear grade cellularity, mitotic rate > 1/50 high power fields (HPF), necrosis and vascular invasion. The clinical behavior of malignant PEComa of the uterus is unknown, but recurrences have been described as well as death from the disease. Therapy currently consists of wide local excision.