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Malignant perivascular epithelioid cell tumor of the uterus with aggressive behaviour: a case report.

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Background: The perivascular epithelioid cell tumor (PEComa) is recognized by the World Health Organization as a mesenchymal neoplasm showing at least partial morphological or immunohistochemical evidence of a putative perivascular epithelioid cell differentiation. This family of tumors includes angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), clear cell sugar tumor of the lung and distinctive clear cell tumors at various other anatomic sites. Occasionally, PEComas are associated with the tuberous sclerosis complex (TCS). Because non-AML/non-LAM PEComas are extremely rare and their nature and prognostic features undefined, we present a case of uterine PEComa with obviously aggressive behavior.

Case Report: A 55-year-old (gravida-2 para-2) presented with a large uterine mass palpated during a routine gynecological examination. The patient did not have any stigmata or family history of TSC, and had no history of melanoma. A transvaginal ultrasound revealed an enlarged uterus with a heterogeneous mass which size was estimated at 9 x 9 cm. The clinical impression of the lesion was a leiomyoma. Total abdominal hysterectomy with bilateral salpingo-oophorectomy and omentectomy was performed 9 months later, and the pathohistologic diagnosis was malignant PEComa of the uterus with metastases in the omentum. Three months after the surgical intervention the patient is still receiving adjuvant chemotherapy and there is no evidence of recurrence.

Results: The uterine body was enlarged and measured 12 x 14 x 8 cm, with lobulated surface. Within the myometrium there was a poorly demarcated gray-tan nodular tumor occupying the fundus, anterior and posterior walls with focally hemorrhagic areas and necrosis on cut surface. The largest diameter of the tumor was 14 cm. In the adipose tissue of the omentum there were numerous nodular tumor masses whose diameters ranged from 1 to 3 cm. Histologically, the tumor demonstrated a tongue-like growth pattern and was composed of large epithelioid cells with clear or eosinophilic cytoplasm, partially arranged around blood vessels. The nuclei were mainly round, centrally located, and vesicular, with moderate to severe pleomorphism and hyperchromatism. Extensive cellular atypia, including bizarre multinucleated giant cells with large nuclei and nucleoli, was present as well. Mitotic count was high (15 per 50 HPFs), with prominent coagulative tumor necrosis. The microscopic examination of the nodular tumor mass from the omentum exhibited the same morphological features. Immunohistochemically, the tumor cells showed positivity for HMB-45 and smooth muscle actin while they were negative for cytokeratins, carcinoembryonic antigen, S-100 protein, CD-10, caldesmon and desmin. Immunoreactivity for vimentin was inconspicuous. The estrogen and progesterone receptors were also negative. A proliferative index of 30% was noted with Ki-67 immunostaining. The morphology and immunohistochemical profile of the lesions was consistent with PEComa.

Conclusions: Due to the fact that a vast majority of PEComas has a benign course of disease or is considered to be of uncertain malignant potential, we present this case with evident aggressive behavior since we believe that the histological features of this tumor can serve as a model for the defining of the criteria for potentially malignant PEComas.

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Prognostic factors in endometrial carcinomas

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Background: The purpose of this study was to present the prognostic factors for endometrial carcinomas. Tumor type, grade of differentiation, depth of invasion and stage have been proposed as prognostic factors and all have been correlated to survival. Bcl-2 (a suppressor of apoptosis) and Ki-67 (a marker of cell proliferation) and mean nuclear volume have also provided prognostic information.

Design: We evaluated histologically 102 specimens according to the recommendations from WHO and FIGO (tumor type, grade of differentiation, depth of invasion and stage). Bcl-2, Ki67, mean nuclear volume and mitotic index were determined using immunohistochemistry and stereology.

Conclusions: The histological parameters, Ki67, mean nuclear volume were significantly correlated to survival and recurrence. Mitotic index was significantly correlated with survival and bcl-2 was neither correlated to survival nor to recurrence.

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Endometrial window of implantation in infertility

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Background: Endometrial window of implantation adequacy plays crucial role in assisted reproductive technologies (ART) success. Therefore morphological and immunohistochemical description of endometrium is significant indicator for prosperous in vitro fertilization and embryo transfer. This study object is evaluation of the endometrial histopathologic, ultrastructural and immunohistochemical features in women applied for ART programs.

Design: Curettages and Pipelle biopsies 21–24 days endometrium from 59 women (26–40 years) with primary (26) and secondary (29) infertility asked for ART treatment were obtained. Twenty-nine patients possessed tubal factor of infertility, 14 - ovulatory factor, 16 - endometriosis. As control were used 10 curettages mid-luteal phase endometrium from healthy women (25–40 years) obtained simultaneously with intrauterine devices removal. Routine histological examination and immunohistochemical determination of estrogen (1D5; «Dako»), progesterone (1A6, «Dako») receptors (ER and PR), Ki67 (MIB 1, «Dako»), P27 (DCS-72.F6, «Diagnostic Biosystems»),

