

# Case report on uterine PEComa: Diagnostic challenges

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## ABSTRACT

**Introduction:** Perivascular epithelioid cell tumour (PEComa) are rare mesenchymal tumours originating from perivascular epithelioid cells. Common sites include gastrointestinal and uterine, with nearly 25% involving uterine corpus. The significance of distinguishing molecular classification governs the benefit from targeted therapy with mTOR inhibitor. **Case Description:** We present a case of a 55-year-old woman with postmenopausal bleeding whom was diagnosed with submucosal fibroid. Transcervical resection of myoma showed symplastic leiomyoma. Hysteroscopy and endometrial tissue sampling revealed smooth muscle tumour of uncertain malignant potential (STUMP) with high suspicion of leiomyosarcoma. The patient underwent completion surgery total abdominal hysterectomy with bilateral salpingo-oophorectomy. The tumour showed a high-grade mesenchymal neoplasm with markers positivity favouring malignant PEComa. **Discussion:** PEComas are rare tumours of female genital tract. Mostly present at fifth to sixth decades of life; signs and symptoms are usually nonspecific. Pathogenesis of PEComa remains unclear. Histopathology and immunohistochemistry play important role in establishing diagnosis. Physical examination of pelvis and pelvic ultrasound frequently yield benign entity; thus, may delay treatment for patients. Surgery remains the mainstay of treatment; while chemotherapy for malignant PEComas aim to reduce the risk of recurrence. Combining chemotherapy and radiotherapy may have a role of increasing disease-free survival. However, there is no uniform treatment regime due to lack of sufficient reports. **Conclusion:** Uterine PEComa diagnosis prove to be a challenge to clinicians by and large. The best diagnostic and management method is yet to be discovered considering the rarity of this neoplasm.