Case Report

Perivascular Epithelioid Cell Tumour of the Uterus: A Case Report and Review of the Literature

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Introduction

Perivascular epithelioid cell tumour (PEComa) is a rare mesenchymal tumour, which is composed of histologically and immunohistochemically distinctive perivascular epithelioid cells [1], which may occur at any anatomic site. In the female genital tract, uterine PEComas are commonest, with 78 reported cases to date [2], in patients aged 17–79 years, with the mean age of 54 years [3]. The vast majority of these tumors are benign or of uncertain malignant potential although rare malignant cases have been reported [4]. We present a rare case of a 56-year old woman with uterine PEComa who presented to our department with postmenopausal bleeding.

Case Report

A 56-year old Caucasian woman presented with history of two episodes of heavy post-menopausal bleeding, requiring hospital admission while on Hormonal Replacement Therapy for five years. She was generally fit and well, with no significant gynaecological history.

Pelvic ultrasound revealed bulky fibroid uterus and a well circumscribed mixed echo solid area within the cervix measuring 3 × 2 cm, consistent with cervical fibroid. The endometrium was 2 mm thin and a trace of fluid was seen within the cavity. The ovaries were not identified and there were no obvious adnexal masses, cysts or free fluid. Outpatient hysteroscopy and endometrial biopsy were performed, which were both normal.

In view of the symptoms, she underwent an uneventful Total Abdominal hysterectomy and bilateral salpingo-oophorectomy. Histology showed normal cervix, fallopian tubes and left ovary. Within the myometrium there was a leiomyoma and a poorly circumscribed irregular nodule composed of nests and cord of epithelioid cells with intervening hyalinised stroma. There was identifiable involvement of lymphovascular channels around the periphery of the lesion. Immunohistochemistry was undertaken with the following results: positive epithelioid cells with desmin smooth muscle actin, vimentin, HMB-45 and SMA. These features were characteristic of a perivascular epithelioid cell tumour (PEComa).

Following discussion at the regional gynaecology oncology multidisciplinary team (MDT) meeting, a Computerized Tomography (CT) scan of the chest, abdomen and pelvis was recommended which did not show any
evidence of malignancy. She remained under close follow up with four monthly appointments in gynaecology outpatient clinic.

**Discussion**

Perivascular epithelioid cell tumours were first described in 1992 and thereafter in 2002 the World Health Organization defined PEComas as mesenchymal tumors composed of perivascular epithelioid cells that express both melanocytic (such as HMB-45, Melan-A) and smooth muscle markers (such as actin and desmin) [1].

The tumour usually affects females [1]. PEComa has been reported to involve multiple organs, such as lung, liver and uterus [5-6]. To date, 78 cases of uterine corpus PEComas have been reported in the English language literature [2], followed by ovarian PEComas with only 1 reported primary case and 6 reports of PEComa metastatic to the ovaries [5].

Uterine PEComas could present with various symptoms, such as abnormal uterine bleeding, postmenopausal bleeding like in our case, uterine mass, pelvic pain and finally as uterine rupture [6]. The behavior of PEComas varies, as some of these tumours could show aggressive behaviours such as metastasis to distant organs, recurrence or death within few years following the diagnosis.

There were not any definite criteria to establish the nature and outcome of women who underwent treatment after being diagnosed with PEComas until 2005, when Folpe et al. proposed risk stratification criteria based on high-risk histopathological findings [1]. These criteria have been applied widely and include large tumour size of more than 5 cm, infiltrative growth pattern, high nuclear grade and cellularity, mitotic rate at least 2 to 3 per 50 HPF and finally the presence of necrosis and lymphovascular invasion.

According to a modification of the Folpe criteria in 2015, tumours of uncertain malignant potential were defined the ones with a tumour size of larger than 5 cm but had no other high-risk features or possessed only the presence of nuclear pleomorphism and multinucleated giant cells. Malignant tumours were defined as those containing any necrosis or at least one high-risk histopathological feature [2].

The differentiation among uterine mesenchymal neoplasms, including leiomyosarcoma, endometrial stromal sarcoma and PEComas can be challenging, therefore both morphologic and immunohistochemical evidence are of the utmost importance for diagnosis [5].

Treatment of PEComas and prognostic factors are not well established in view of their rarity. Hysterectomy remains the standard treatment of choice, despite the fact that long term outcomes are still unclear. Their malignancy potential is unpredictable, therefore close follow up is essential. In patients with tumours demonstrating malignant behaviours, surgical resection is the gold standard, followed by possible radiation and cytotoxic chemotherapy in both neoadjuvant and adjuvant therapies [1]. Our patient had no any symptoms of recurrence after the operation, however she is on long-term follow up.

In conclusion, uterine PEComas are rare and should be included in the differential diagnosis of mesenchymal tumours, as their differentiation can be challenging. Their diagnosis is essentially based on examination of morphologic features and immunohistochemistry. Early suspicion of abnormal myometrial tissue and appropriate treatment could lead to a better outcome for patients.

**Conflicts of Interest**

Dr. Efterpi Tingi and Dr. Khawla Aswad declare that there has been no conflict of interest.
References


