Case Report

Perivascular Epithelioid Cell Tumor (PECom) of Vagina: A Rare Occurrence - ⚗

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INTRODUCTION

Perivascular epithelioid cell tumor (PEComa) is a rare subtype of mesenchymal origin tumors, and is composed of perivascular epithelioid cells with specific histologic and immunohistochemical features [1]. PEComas can occur at any anatomic site and include angiomyolipomas, lymphangioleiomyomatosis, clear cell “sugar” tumors of the lung, and PEComa not otherwise specified [2,3]. PEComa of the female gynecological tract is a rare entity. When considering PEComas of the female genital tract, the uterus is the most common location. Involvement of the ovary in the context of a primary uterine PEComa, in the absence of systemic disease associated with tuberous sclerosis, however, has only been reported in 1 previous case [4]. One case of primary PEComa of vagina was reported from china [5]. Second case was reported in a eight year old girl [6].

CASE REPORT

Our patient was a 36-year-old woman who presented with vaginal mass. Her past medical history was unremarkable. Initial findings were a 2/2 cm of vaginal mass on pelvic examination. A punch biopsy of vaginal mass was performed. Histologic and immunohistochemical findings provided the most accurate means of diagnosis [7]. Although PEComas are often benign, there have been reported cases of malignant tumors. To distinguish between benign and malignant uterine PEComas, the Folpe criteria are often used [8]. According to the Folpe criteria, a tumor is considered malignant if it contains at least 2 worrisome features, defined as size of at least 5 cm, high nuclear grade and cellularity, a mitotic rate of at least 1 per 50 HPF and necrosis or vascular invasion. In 2015, a modification of the Folpe criteria was proposed, defining malignant tumors as those containing any necrosis or at least 1 worrisome feature, defined as an invasive edge, size of at least 5 cm, a mitotic rate of at least 2 to 3 per 50 HPF, and lymphovascular invasion [9].

CONCLUSION

Distinguishing among mesenchymal neoplasms, including PEComas, endometrial stromal sarcomas, and leiomyosarcomas, can be difficult. Careful analysis of morphologic and immunohistochemical features is of the utmost importance.

REFERENCES