with no or minor areas of peripheral infiltration, or display a prominent "tongue-like" infiltration into the myometrium. Multinucleated giant cells, which appear to be degenerative in nature may also be found. PEComas may display diffuse stromal hyalinization as to obscure their underlying features. Rare features include sex cord like features[17] and prolactin secretion[18]. Sclerosing PEComas also have been described[19]. Froio et al. described a case of multifocal PEComa of the female genital tract associated with endometriosis, diffuse adenomyosis, and endometrial atypical hyperplasia[20].

On immunohistochemistry, PEComas co-express melanocytic and smooth muscle markers. Melanocytic markers such as HMB-45, HMSA-1, MelanA/ Mart1, microophtalmia-associated transcription factor (MiTF) are positive with HMB-45 found to be the most common melanocytic marker of uterine PEComas, with 99% positivity[21]. Additionally, SMA is considered to be the most common smooth muscle marker, with positive results in 80% of uterine PEComas, with less immunoreactivity for desmin and caldesmon[22]. Vimentin is usually inconspicuous in these tumors. The expression of these markers may vary as predominantly epithelioid pattern may strongly express melanocytic markers and limited myoid marker, while predominantly spindle cell pattern shows reverse expression. Recently, Cathepsin K has emerged as a sensitive marker for PEComas, although non-specific, being also expressed in leiomyosarcoma, melanoma, and alveolar soft part sarcoma[23,24]. A new entity, TFE3 translocation associated uterine PEComas have recently shown positive immunoreactivity for HMB-45, TFE3, and Cathepsin K and negativity for MiTf, SMA and desmin[25].

The distinction between common differential diagnosis of mesenchymal uterine neoplasms, including leiomyosarcomas, PEComas, and endometrial stromal tumors is often diagnostically challenging. Morphologic features such as the identification of a capillary network are of paramount importance in distinguishing between mesenchymal neoplasms. Leiomyosarcomas lack a capillary network, and although a capillary network is often present in ESS, the capillaries lack the radial appearance of epithelioid cells characteristic of PEComas. Because of their expression of HMB-45 and/or melan-A, PEComas are frequently confused with both conventional melanoma and clear cell sarcoma. Some of the morphologic features of PEComas, including the admixture of spindled and epithelioid forms, the occasionally prominent nucleoli, and the presence of multinucleated cells, are also seen in melanoma and clear cell sarcoma. In most cases, melanoma/clear cell sarcoma can be distinguished from PEComa by their strong expression of S-100 protein and smooth muscle actin non-immunoreactivity. Although morphologic features may overlap, use of immunohistochemistry helps to diagnose challenging cases. PEComas are often benign with few cases of malignant tumors being reported. To distinguish between benign and malignant uterine PEComas, the Folpe criteria are often used[26]. 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