of the Folpe criteria defined 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. Thus extensive sampling is warranted to look for worrisome features thereby affecting prognosis of the patient. The most common sites of metastasis of PEComas are lung, followed by liver, lymph nodes, and peritoneum. PEComa rarely metastasize to the ovaries, the majority of which have been associated with multiorgan involvement in the context of tuberous sclerosis.

There is no established treatment protocol for uterine PEComas. At present, complete resection is the preferred treatment modality for PEComa, but long-term outcomes remain unclear. Hysterectomy and adnexectomy are considered as the most direct and effective means of treatment. But for the malignant PEComa of uterus, pelvic lymph nodes dissection and postoperative adjuvant chemotherapy remains controversial. The plan of chemotherapy is still under research. As a result, close follow-up is necessary for all patients diagnosed with PEComas and is especially important for those diagnosed with PEComas of malignant potential. Our case underwent panhysterectomy with uneventful postoperative period and close follow up. PEComas are a rare group of neoplasms that are difficult to differentiate from other uterine epithelioid and mesenchymal tumors on clinicoradiologic features. Careful examination of morphologic features and immunohistochemistry are necessary to arrive at an accurate diagnosis. The pathologist should always consider the possibility of a rare neoplasm such as PEComa in the list of differentials thus emphasizing the need of extensive sampling and use of appropriate immunohistochemical studies, wherever necessary.

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