FINAL DIAGNOSIS: Perivascular Epithelioid Tumor (PEComa) of the uterus.

DISCUSSION

In recent years, a group of tumors that have been designated "perivascular epithelioid cell tumors" (PEComa) have been reported with increasing frequency from a wide variety of anatomic locations. The term PEComa was first introduced in 1996 by Zamboni et al to describe one such case arising in the pancreas[1]. PEComas belong to an identically named family of tumors comprised of conventional angiomyolipomas, clear cell sugar tumors, lymphangiomyomatosis, and clear cell myomelanocytic tumor of the falciform ligament/ligament teres. The cell of origin of PEComas is still controversial, yet embryological and in vitro studies have shown evidence for origin from a neural crest stem cell that is capable of both myoid/myogenic and melanocytic differentiation[2]. PEComas display a striking female preponderance and appear to be anatomically ubiquitous[3,4]. In the gynecologic tract, the uterus is the most common location for PEComas, with 78 reported cases till date[5-12]. The age of these patients vary from 9 years to 79 years (mean = 45 years). The clinical presentations are diverse and vague, and include abnormal vaginal bleeding, abdomino-pelvic pain, uterine rupture, and hemoperitoneum; being incidental findings in some cases[3,13]. Uterine PEComas have also been associated with tuberous sclerosis (9%)[14]. A rare case of uterine PEComa with situs inversus was reported by Han et al[15].

The radiologic appearances have been similarly variable, may be small and homogeneous, simulating a benign smooth muscle neoplasm or they may be large, lobulated, and heterogeneous masses[16]. Thereby, neither the clinical presentation nor radiologic appearance of uterine PEComas is sufficiently distinctive to allow the diagnosis to be suggested preoperatively.

PEComas usually present as a solitary or rarely multifocal mass in the uterine corpus from 0.6 cm to 12.0 cm (mean = 4.7 cm). The cut sections may be tan-white or tan-grey or grey-white, with whorling, soft and fleshy texture, and ill-defined, or rarely circumscribed margins. On microscopy, the tumor is characterized by a diffuse, nested, and/or fascicular proliferation of epithelioid-looking cells that display clear, granular, or eosinophilic cytoplasm with little intervening stroma. The purely spindled and admixture of spindle and epithelioid patterns are also described. Mitotic activity is usually low, around 0-1 per 50 hpf and may show varying degree of nuclear pleomorphism, macronucleoli, intranuclear psuedoinclusions, or “spider cells”[7]. The constituent cells may display a perivascular distribution. PEComas typically display a prominent network of small capillaries reminiscent of renal clear cell carcinoma or myxoid liposarcoma. The tumors may be circumscribed