

A 44 Year Old Woman With Retroperitoneal Mass: A Pathology Quiz

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Citation

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Abstract

PATHOLOGY QUIZ

A 44-year-old woman presented with complaint of right-sided back pain for one month. MRI and CT scans, performed at an outside institution, revealed a 10 cm retroperitoneal mass posterior to the right kidney, pushing it anteriorly. The mass was not connected to the kidney, adrenal gland, vascular or any other structures. Plasma and urinary metanephrine were within normal limits. Past history was significant for a high-grade ductal carcinoma in-situ of left breast s/p lumpectomy and a total abdominal hysterectomy with bilateral salpingo-oophorectomy for leiomyomata (both procedures were done two years prior). CT-guided biopsy of the mass was non-diagnostic.

Subsequently, the patient underwent surgical excision of the mass. Grossly, the resected specimen consisted of a well-circumscribed encapsulated firm ovoid gray-tan mass measuring 14 x 10 x 6 cm, with attached fibrofatty tissue. Serial sections revealed a lobulated gray homogeneous cut surface (Figure 1). A cystic area measuring 4 cm was present towards one periphery of the mass.

Microscopically, the tumor consisted of myoid spindle cell proliferation arranged in irregular sheets and nests separated by prominent stromal hyalinization. The tumor cells displayed vacuolated to eosinophilic cytoplasm with pink globules (Figure 2A). Areas of dense hypercellularity and fascicular arrangement were present (Figure 2B). Radial and concentric arrangement of tumor cells was prominent around thick-walled medium sized malformed blood vessels (Figure 3). Nuclear pleomorphism was mild and mitosis were rare (<1/50 HPF). No tumor necrosis, vascular invasion or infiltrative growth pattern were seen.

The tumor cells showed diffuse positivity for desmin and smooth muscle actin (Figure 4A & B) and scattered

positivity for HMB-45 and S-100 protein (Figure 4C & D). CD34 highlighted the vascular endothelial lining but tumor cells were negative. No immunoreactivity was noted for Melan-A, CD99, CD10, inhibin, calretinin and pancytokeratin (CKAE1/AE3).

Figure 1

Figure 1: Gross appearance of the tumor cut surface.

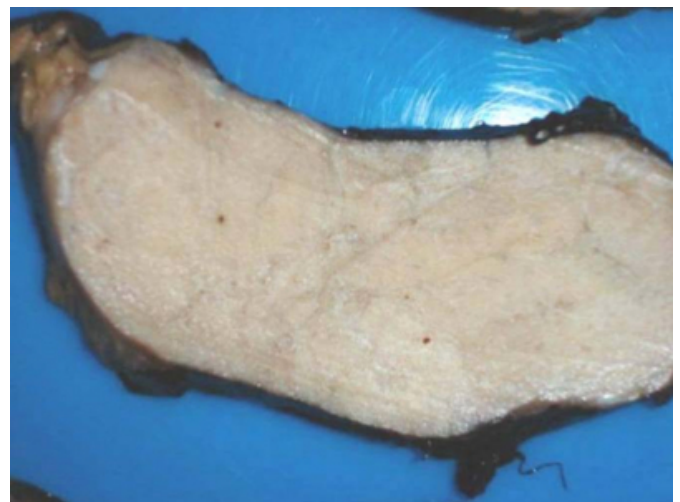


Figure 2

Figure 2: Microscopic pictures representative of the tumor (H&E x 200).

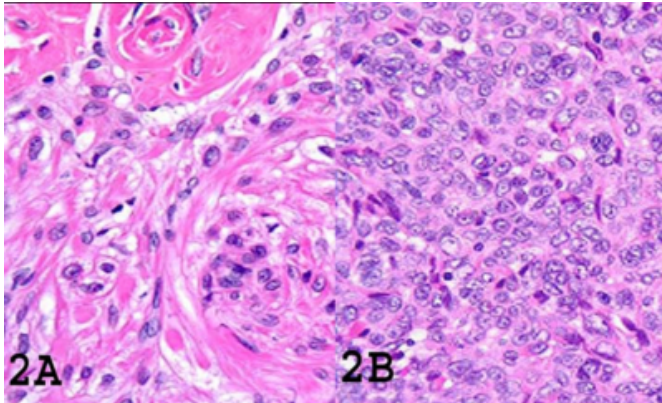


Figure 3

Figure 3: Concentric arrangement of tumor cells around thick-walled malformed blood vessels (H&E x 200).

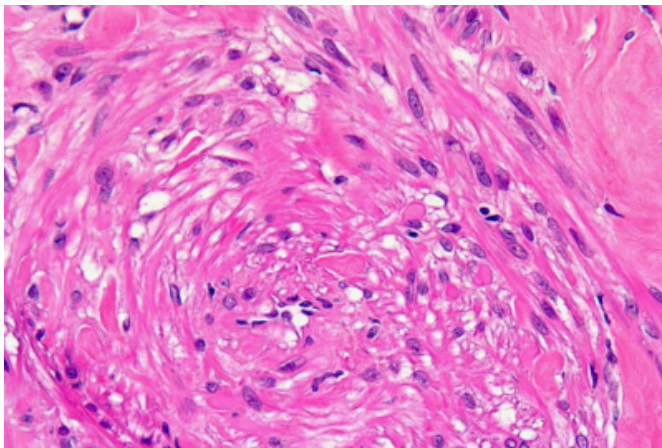
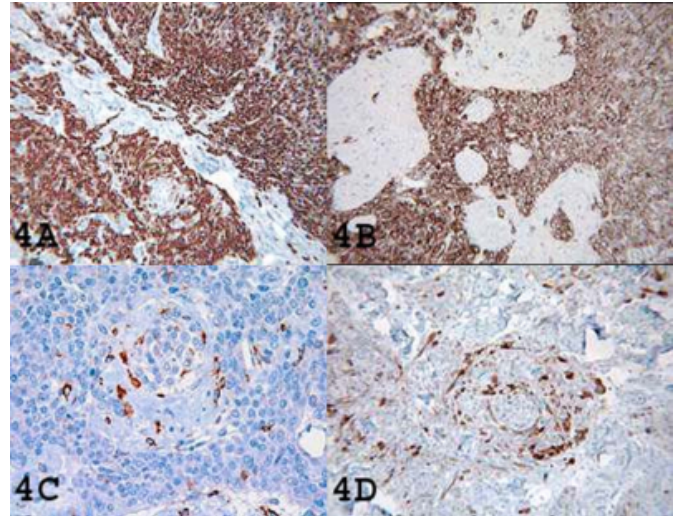


Figure 4

Figure 4: Immunostains (A: Desmin; B: Smooth muscle actin; C: HMB-45; D: S-100 x 100).



WHAT IS YOUR DIAGNOSIS?

Pathologic Diagnosis: PEComa (tumor showing perivascular epithelioid differentiation) with features favoring angiomyolipoma.

We report a case of PEComa with features of angiomyolipoma in a 44-year-old woman presenting as a retroperitoneal mass posterior to the right kidney. Grossly, the tumor was 14 x 10 x 6 cm, well circumscribed and firm, with a lobulated gray tan homogenous cut surface and focal area of cystic change. Microscopically, the tumor was formed of myoid spindle cells with radial and concentric arrangement around thick-walled malformed blood vessels. Smooth muscle and vascular components were dominant in the tumor with absence of lipomatous component despite extensive sampling. Areas of dense hypercellularity with fascicular pattern were focally present. The tumor immunopositivity for melanocytic (HMB-45, S-100) and smooth muscle markers (desmin, smooth muscle actin) was typical for PEComa. The tumor histomorphology and absence of immunoreactivity for pancytokeratin (CKAE1/AE3), CD99, CD10, inhibin, and calretinin excluded desmoplastic small round cell tumor, adrenocortical tumor, renal cell carcinoma and mesothelioma.

PEComas are a family of related mesenchymal tumors distinctive for “perivascular epithelioid cells” and immunoreactivity to both melanocytic and smooth muscle markers.[1] The PEComa group of tumors includes angiomyolipoma (AML), lymphangiomyomatosis, clear cell

“sugar” tumor of lung, and rare similar tumors reported in a variety of visceral and soft tissue sites.^[12]

Angiomyolipoma most commonly arises in the kidney. It may also occur in the liver and very rarely in other anatomical sites such as retroperitoneum, nasal cavity, oral cavity, heart, colon, lung, and skin.^[3,4,5,6,7,8,9,10]

In the classic form of AML a triphasic admixture of smooth muscle cells, thick walled vessels and adipose tissue are present, but the relative proportion of different components is variable. Some cases predominantly exhibit only one or two tissue components, rare cases are exclusively formed of only one component such as the monotypic epithelioid variant of AML.^[11] Our case showed biphasic angiomyomatous components with no detectable lipomatous element.

Angiomyolipoma can be associated with several hereditary disorders including tuberous sclerosis complex (TSC), Von Hippel Lindau syndrome, autosomal dominant (adult type) polycystic kidney disease and Von Recklinghausen disease. Among these hereditary disorders, TSC shows the highest association with renal AML.^[3] TSC is a group of autosomal dominant genetic disorders caused by germ-line mutation at the chromosomes 9q34 and 16p13.3 in the genes encoding for the protein hamartin (TSC1 gene) and tuberin (TSC2 gene) respectively. About 80% of patients with TSC are affected by AML, which can be multiple and bilateral in these patients. Almost half of patients with renal AML present with manifestations of TSC.^[12] The association of TSC with hepatic AML is less frequent being present in only 5-10% of patients.^[3]

The criteria of malignancy or those predictive of aggressive tumor behavior are not yet well defined for AML due to the rarity of these lesions. The data from the rare aggressive cases associated with metastases and death suggested that adverse features include any combination of tumor size >8 cm, infiltrative growth, marked hypercellularity, high nuclear grade, mitotic activity >1 mitosis/50HPF, atypical

mitosis and coagulative necrosis.^[12] However, metastases were also reported from PEComas that showed no histological features of malignancy.^[13] This case was considered to be of “uncertain malignant potential”, and close follow-up of the patient was recommended due to the large size of tumor (14 cm) and presence of focal areas of hypercellularity.

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