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Case Report

Sclerosing PEComa: A Histologic Surprise

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ABSTRACT

PEComa represents a group of mesenchymal tumors consisting of perivascular epithelioid cells. We present a 50-year old female patient with a rare distinctive variant, sclerosing PEComa, characterized by extensive stromal hyalinization and a predilection for the pararenal retroperitoneum.

Key Words: PEComa, Sclerosing PEComa

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INTRODUCTION

Sclerosing PEComas are rarely encountered in clinical practice. Most reported cases follow a benign course. Currently, surgery is the mainstay of treatment[1]. There is a paucity of data regarding diagnostic criteria, prognostic factors and management options. We report a case of sclerosing PEComa arising from the right pararenal space.

CASE REPORT

A 50 year old female patient presented with right loin discomfort of one month duration. She had no comorbidities. General and systemic examination revealed no abnormality other than pallor. Urine microscopy was normal. Hematological and biochemistry investigations revealed anemia (Hb 8.4mg/dl). Ultrasonography showed an 8×8 cm mass arising from the right kidney. Contrast enhanced CT scan confirmed a well-defined, rounded, exophytic, heterogeneously enhancing lesion arising from the midpole of the right kidney, located in the right pararenal space (Fig. 1).

The patient received 2 units packed red cells preoperatively and underwent an uneventful right radical nephrectomy via an 11th rib extraperitoneal approach. An 8×8cm exophytic lesion was seen arising from the medial aspect of the right kidney, abutting the duodenum medially. There was no evidence of regional lymph node enlargement or tumor thrombus.

PATHOLOGICAL FINDINGS

A well encapsulated pararenal mass measuring 8×7×6 cm was seen arising from the hilar area of the right kidney indenting the renal parenchyma. It was solid, firm, with a homogenous grey white surface (Fig. 2).

Histologically, the tumor was composed of a dimorphous population of cells. One component was of uniform bland cells, small to medium sized, with faint granular to clear cytoplasm. Nuclei were round with indistinct nucleoli. These cells were interspersed with extensively hyalinised capillaries.

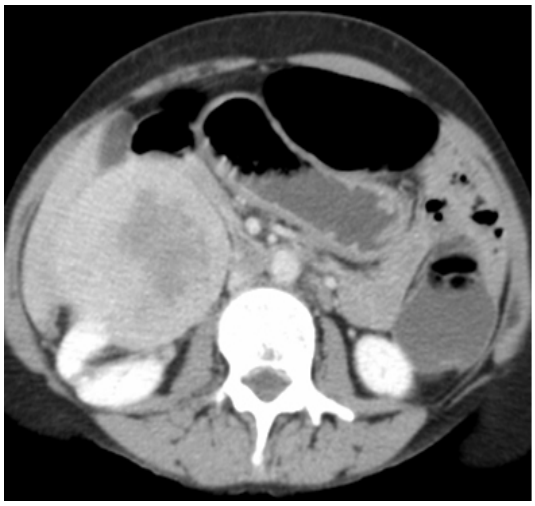

Fig. 1: Contrast enhanced CT image–axial section demonstrating a well-defined, rounded, exophytic, heterogeneously enhancing lesion seen arising from the midpole of the right kidney, located in the right pararenal space.

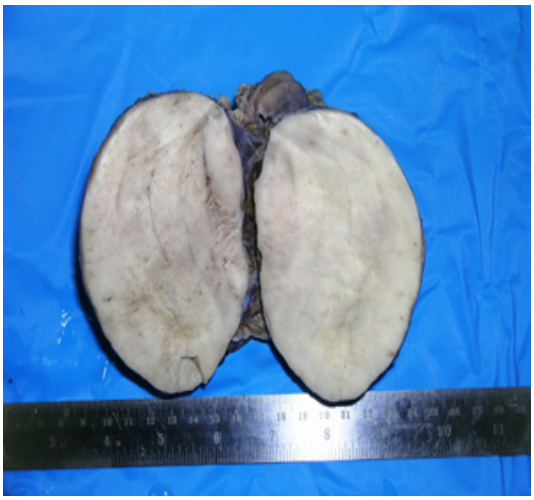

Fig. 2: Gross picture of the operative specimen showing a well encapsulated pararenal mass.

A second component of fascicles of spindle cells, closely intermingled with the round cells was present. Large vascular channels with both round and spindle cells radiating from the walls were seen (Fig. 3). Mitotic activity was 1/50 HPF. Cells were embedded in an abundant sclerotic stroma (Fig. 4).

Immunohistochemistry revealed diffuse positivity for smooth muscle actin (cytoplasmic and membranous) (Fig. 5), HMB 45 (cytoplasm) was strongly positive in perivascular cells (Fig. 6).

CD99 and BCL2 (cytoplasmic and membranous) were positive. CK, CK7, CD117 and synaptophysin were negative. MIB1 (proliferation index) was 1/100 cells. All antibodies (Dako) were in ready to use form. Antigen retrieval was done with a pressure cooker (2 minutes).

DISCUSSION

PEComas are a family of mesenchymal neoplasms that include angiomyolipma, clear cell sugar tumor of the lung and lymophangiomyomatosis[2]. These tumors, which show perivascular epithelioid differentiation, are composed of PEC cells (perivascular epithelioid cells) which have no normal counterpart. These cells were described by Bonneti et al who noted their association with blood vessels and coined the term PEComas[3].

PEComas are generally composed of epithelioid cells with clear to granular pale eosinophilic cytoplasm. Less often they are accompanied by a spindle cell component. Immunohistochemically, they show a mixed smooth muscle and melanocytic phenotype. Our case is similar to the conventional PEComa but had a sclerotic stroma, which is described in sclerosing PEComas which are a distinctive variant with a predilection for the retroperitoneum of middle aged women. They are known to lack the delicate vasculature of the conventional PEComa[2].

Sclerosing PEComas usually pursue an indolent course. Folpe et al[4] recommend that any combination of the following features warrants a diagnosis of malignancy: Large tumor size (greater than 5cm), mitotic activity >1/50 HPF, necrosis, high nuclear grade and cellularity and infiltrative growth. A diagnosis of "uncertain malignant potential" is justified if only one of these features

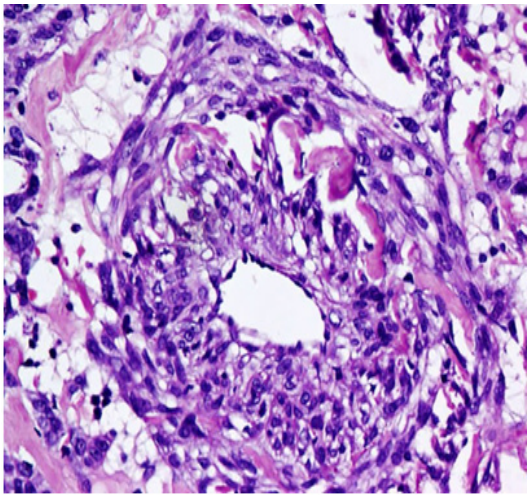

Fig. 3: Photomicrograph showing large vascular channels with both round and spindle cells radiating from the walls (x150).

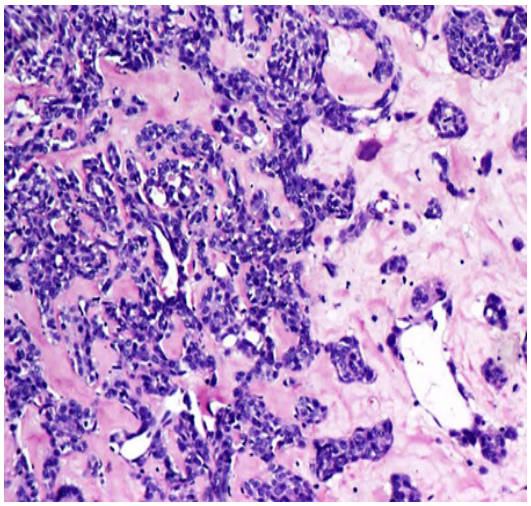

Fig. 4: Photomicrograph showing cells embedded in a sclerotic stroma (x100).

is noted. On account of tumor size (8cm) the tumor in our patient was designated to be of uncertain malignant potential.

The chief differential diagnostic considerations in this case were synovial sarcoma, epithelioid smooth muscle tumors and glomangioma.

Synovial sarcomas are characterized by brisk mitotic activity. While these tumors are reactive for BCL2 and frequently reactive for CD99, they are negative for smooth muscle actin[5]. This tumor was negative for CK and CK7. CD99 and BCL2 positivity are documented in PEComas[6].

Like smooth muscle tumors, sclerosing PEComas express smooth muscle actin, but unlike them, they express HMB-45 which helps in distinguishing these tumors[2]. The clear to pale cytoplasm and the relationship with blood vessels that are characteristic of PEComas are helpful in distinguishing them from epithelioid smooth muscle tumors[2].

Cell morphology and the perivascular orientation suggests a diagnosis of glomangioma, but these tumors are known to occur in the extremities and are associated with cavernous vascular channels[7]. Gastrointestinal stromal tumor and carcinoid were ruled out based on morphology and negative staining for CD117 and synaptophysin, respectively.

The optimal treatment for PEComas is not well established. Currently, surgery is the mainstay of treatment for primary PEComa at presentation as well as for local recurrences and metastases, with the aim of obtaining clear resection margins[1, 6]. Metastases have been successfully managed by resection alone. Primary excision is usually curative, as most PEComas are benign. Adjuvant therapies, including chemotherapy and immunotherapy, may be considered for patients with locally advanced or metastatic PEComa. Of particular interest is the efficacy of the mTOR inhibitor rapamycin in animal model studies[9].

To summarize, sclerosing PEComa is an unusual tumor that has a predilection for the retroperitoneum in middle aged women. It is analogous to the sclerosing variant of the epithelioid angiomyolipoma[8].

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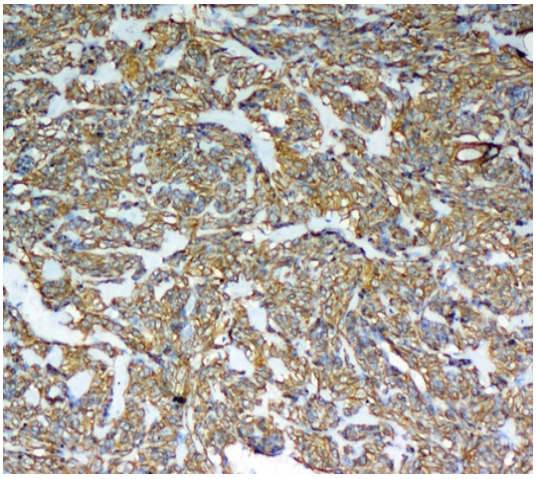

Fig. 5: Photomicrograph showing positivity for smooth muscle actin (x100).

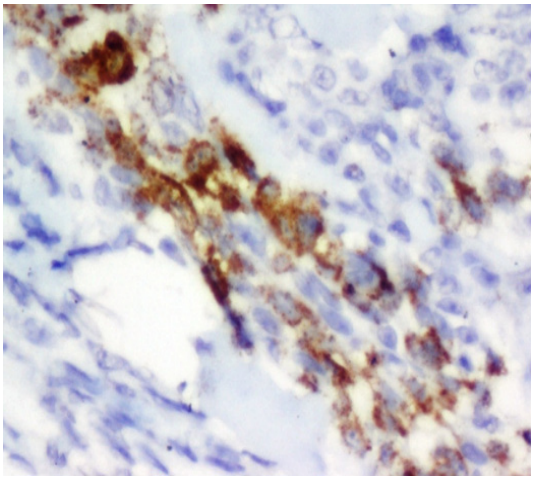

Fig. 6: Photomicrograph showing positivity for HMB45 (x400)

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