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Renal PEComa in a young male: A case report and insights from the literature

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ABSTRACT

Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal neoplasms, commonly found in the uterus and retroperitoneum. Renal PEComas are exceedingly rare, often posing diagnostic challenges due to their resemblance to renal cell carcinoma (RCC) on imaging. We present the case of an 18-year-old male who presented with non-specific symptoms of fever, chills, and shivering. Imaging revealed a renal mass, initially suspected to be RCC. However, following a right radical nephrectomy, histopathological examination confirmed the diagnosis of malignant PEComa. This case highlights the rarity of renal PEComa and underscores the importance of early diagnosis and appropriate surgical management to prevent complications.

1. Introduction

Perivascular epithelioid cell tumors (PEComas) are a rare group of mesenchymal neoplasms characterized by perivascular cells, with the most common sites being the uterus and retroperitoneum.[1]. PEComas encompass a diverse spectrum of tumors, including angiomyolipoma, clear cell "sugar" tumor of the lung, lymphangioleiomyomatosis, and several atypical tumors located in the visceral organs, abdomen, and soft tissues/bones, such as clear cell tumors.[2]. These neoplasms consist of epithelioid or spindle cells, identifiable through histological and immunohistochemical analysis.[3]. Imaging modalities are often inconclusive, making definitive diagnosis challenging and typically reliant on post-surgical findings. While the majority of PEComas are benign and predominantly affect women,[4], renal PEComas are exceptionally rare. This paper presents a case of renal PEComa, incidentally detected in an asymptomatic young male, and includes a review of previously reported cases due to its rarity.

2. Case report

An 18-year-old male patient presented to our hospital with new onset complaints of fever, chills, and shivering. He had no history of hereditary or infectious diseases, nor did he consume alcohol or smoke. During the physical examination, a mass was detected in the right kidney. Blood analysis results were as follows: hemoglobin 131 g/L, platelet count 294 x 10^3/μL, leukocyte count 8.33 x 10^3/μL, erythrocyte count 5.02 x 10^6/μL, and a normal coagulogram. Biochemical analysis revealed glucose levels of 102 mg/dL, creatinine 0.7 mg/dL, and C-reactive protein 1.3 mg/L.

Seven years prior, a contrast-enhanced computed tomography (CT) scan had revealed the presence of an angiomyolipoma nodule in the lateral region of the right kidney, located in the middle third of the cortical layer. Recent CT and magnetic resonance imaging (MRI) identified several angiomyolipomas, the largest measuring 15× 14 mm, on both kidneys. Additionally, a solid mass measuring 38 x 36 × 35 mm was detected in the middle portion of the right kidney (Fig. 1). Contrast-enhanced imaging showed that the tumor appeared hypovascular compared to the renal parenchyma, with no detectable "washout" effect.

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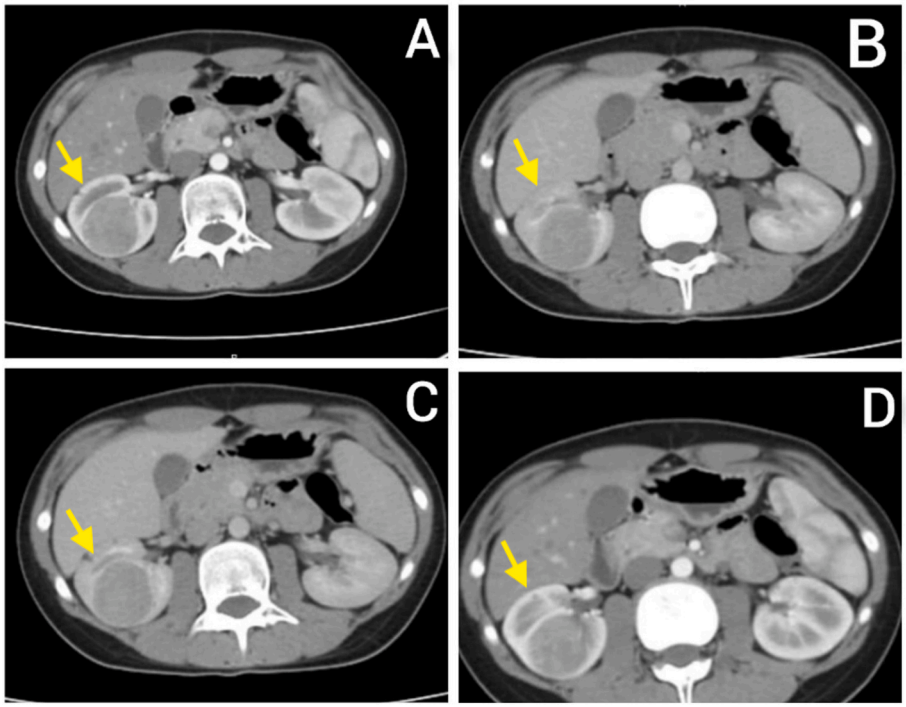

Fig. 1. Right renal mass appearance on abdomen CT scan.

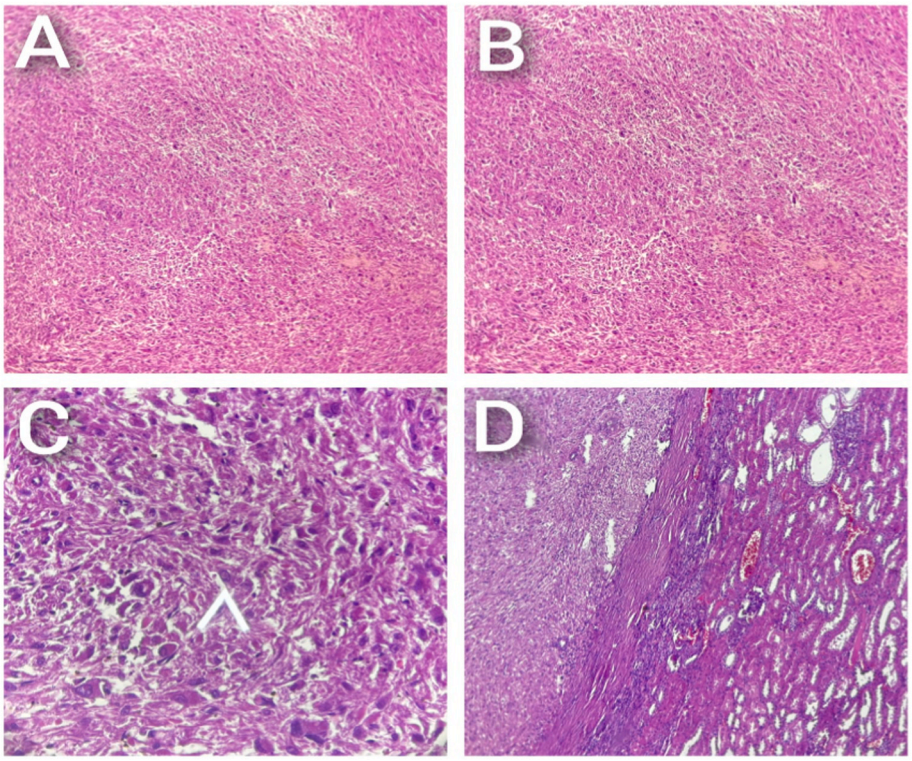

Fig. 2. Histopathology examination demonstrates the presence of capsule invasion, eosinophilic cytoplasm, pleomorphic and atypical cells, occasionally vesicular nuclei and multinucleated giant cells, rhabdoid-like cells, intranuclear pseudoinclusions, and necrosis.

MRI images demonstrated restricted diffusion within the tumor, along with fatty tissue presence, leading to a preliminary diagnosis of renal cell carcinoma (RCC). The patient's symptoms initially suggested a renal abscess, prompting a shift from biopsy to curative treatment.

The patient was subsequently undewent a right radical nephrectomy. Histological examination of the specimen revealed capsule invasion, infiltrative growth pattern, high nuclear grade and cellularity, vascular invasion, necrosis, and high mitotic rate, leading to a diagnosis of malignant PEComa (Figs. 2 and 3).

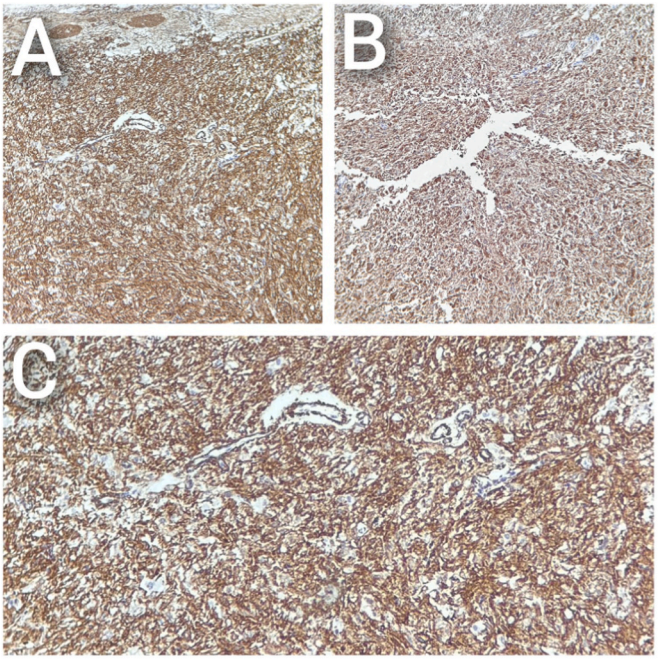

Fig. 3. Immunohistochemical examination showed positive staining for HMB45 (A), α-SMA (B), and S100 (C).

3. Discussion

Renal angiomyolipomas are benign tumors of the kidney, classified within the PEComa group. Accurate diagnosis requires a high degree of clinical suspicion and comprehensive immunohistochemical analysis. From a radiological perspective, PEComas pose a diagnostic challenge due to their similarity to RCC on CT or MRI.

The presenting symptoms of renal PEComa are typically nonspecific and may include back pain, fever, or hematuria. For instance, a 39-year-old male patient presented with new-onset abdominal pain and left flank discomfort, associated with anemia.[5]. Imaging revealed a 30 × 28 cm left renal mass with internal areas of varying density, suggestive of a hematoma. In another case,[6], a 59-year-old male patient with a history of deep vein thrombosis presented with nonspecific abdominal pain. A CT scan showed a 5 × 4 cm contrast-enhancing exophytic mass in the left upper pole of the kidney, without a fatty component. This patient subsequently underwent partial nephrectomy.

Unlike these cases, our patient was asymptomatic. Despite the absence of symptoms, the condition was complicated by the presence of a hematoma, underscoring the importance of early diagnosis and intervention. While the disease course was mild in our case, more aggressive forms have been reported in the literature. In a small subset of patients, tuberous sclerosis, recurrence, and metastasis have been observed, with metastases most commonly occurring in the liver, lungs, and mesentery. Nese et al.[7] identified several negative prognostic factors, including tumor necrosis greater than 7 cm, tuberous sclerosis, extrarenal spread, renal vein invasion, and a carcinoma-like growth pattern.

In another report,[8], a 46-year-old female patient presented with right flank pain, nausea, and vomiting that persisted for two weeks. All laboratory findings were normal, except for elevated fasting blood glucose levels (11.79 mmol/L). Based on the tumor's size and anatomical features, nephron-sparing surgery was performed, and the mass was fully resected. No complications were noted during the one-year follow-up period. In our case, total nephrectomy was performed without postoperative complications, and the patient remains under routine follow-up.

In conclusion, we have identified a rare case of renal PEComa. The gold standard for diagnosis relies primarily on a combination of histopathological and immunohistochemical evaluation. Both nephron-sparing surgery and total nephrectomy are effective treatment options, depending on the tumor's characteristics and location. While the clinical course is often asymptomatic, late-stage complications and metastasis may occur, highlighting the importance of early diagnosis and appropriate surgical intervention.

CRediT authorship contribution statement

Rashad Sholan: Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Software, Resources, Project administration, Methodology, Investigation, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Rufat Aliyev:** Validation, Supervision. **Malahat Sultan:** Writing – original draft, Validation, Supervision. **Anar Almazkhanli:** Writing – original draft, Resources. **Savalan Karim:** Writing – original draft, Visualization, Resources. **Jalal Gasimov:** Writing – original draft, Visualization.

Informed consent

The patient provided informed consent for the publication of their medical information and treatment details. All efforts were made to maintain patient confidentiality, in line with institutional ethical standards.

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