Case Report

Metastatic alveolar soft part sarcoma of the kidney in a young female

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ABSTRACT

A 19-year-old female presented with left flank discomfort and swelling. Imaging revealed a large mass arising from the left kidney, and radical nephrectomy confirmed the diagnosis of alveolar soft part sarcoma (ASPS) based on histopathological and ultrastructural examination. Postoperatively, positron emission tomography–computerized tomography showed lung metastasis and renal bed recurrence. Sunitinib was initiated for metastatic ASPS. This case underscores challenges in diagnosing and managing ASPS, highlighting the role of tyrosine kinase inhibitors. Multidisciplinary care and vigilant follow-up are crucial for rare tumors such as ASPS.

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is an extremely rare malignant tumor that predominantly affects adolescents and young adults.[1] ASPS accounts for <1% of all soft-tissue sarcomas, and its primary occurrence in the kidney is quite rare, with only a limited number of cases reported in the literature.[2] ASPS of the kidney presents a diagnostic challenge due to its nonspecific clinical presentation and radiological features that can resemble other renal neoplasms. In this case report, we present a case of primary ASPS of the kidney in a young female who was diagnosed with metastasis postoperatively.

CASE REPORT

A 19-year-old female patient presented with a 1-year history of left flank discomfort, accompanied by a progressively enlarging swelling in the left flank. The patient did not report hematuria, urinary symptoms, or other systemic complaints. Ultrasonography, chest X-ray, and contrast computerized tomography (CT) scan of the abdomen and pelvis revealed a large, enhancing irregular mass of size 15.9 cm × 15.1 cm × 16.2 cm arising from the upper pole of the left kidney, with compression of the pancreas and involvement of the renal sinus [Figure 1]. Subsequent laboratory investigations ruled out hormonal abnormalities related to the adrenal gland. The patient underwent left open radical nephrectomy, during which a large renal mass measuring 12 cm × 15 cm was identified. Intraoperative findings demonstrated preserved planes and dilated splenic veins with collateral formation. Postoperatively, the patient developed a pancreatic fistula, which was managed conservatively.

Histopathology and immunohistochemistry

Gross examination revealed a large tumor measuring 16 cm × 14 cm × 11 cm and replacing the upper and mid pole of the kidney. The outer surface was bosselated; however, the tumor was not breaching the renal capsule. Cut surface showed large areas of necrosis and hemorrhage. The tumor was infiltrating the renal sinus. Microscopic examination showed an unencapsulated tumor with significant areas of dystrophic calcification in the periphery. The tumor cells

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were predominantly arranged in nested and alveolar patterns. On higher magnification, the tumor cells were large and polygonal with sharply demarcated cell borders, abundant granular eosinophilic cytoplasm, vesicular chromatin, and prominent nucleoli. Periodic acid–Schiff (PAS) stain demonstrated fine granular cytoplasmic positivity that disappeared with diastase treatment in most of the tumor cells. Few of the cells demonstrated scattered PAS-positive, diastase-resistant cytoplasmic granules [Figure 2]. Based on morphological features, possibilities of cellular angiomyolipoma, renal cell carcinoma, adrenocortical carcinoma, and ASPS were considered.

Tumor cells showed immunopositivity for TFE3 and cathepsin K, while other markers such as pan-cytokeratin (CK), epithelial membrane antigen, HMB45, melan-A, inhibin, PAX8, CA-IX, calretinin, CK7, CK20, CD117, S100, AMACR, MITF, and ALK were negative [Figure 3]. SDH-B immunoexpression was retained. Ultrastructural examination revealed well-developed smooth endoplasmic reticulum, prominent Golgi apparatus, occasional neurosecretory granules, and numerous needles to rhomboid-shaped crystalline structures [Figure 3]. Based on these findings, a diagnosis of ASPS was established.

Treatment and follow-up

Postoperatively, the patient developed pancreatic fistula, which was managed conservatively. A positron emission tomography-CT (PET-CT) scan was performed at 3 months, which revealed metastasis to the left lung, not seen during initial evaluation, along with recurrence in the left renal bed. Subsequently, the patient was started on sunitinib, a tyrosine kinase inhibitor. She is currently under follow-up, and further imaging and clinical evaluations will be conducted to assess the response to treatment and disease progression.

DISCUSSION

ASPS is an exceedingly rare malignancy, predominantly affecting children and young adults. While ASPS commonly arises in soft tissues, its occurrence in the kidney is extremely rare, with only a limited number of cases reported in the literature.[2]

The clinical presentation of ASPS of the kidney can be nonspecific, often mimicking other renal neoplasms. In our case, the patient did not have hematuria or urinary symptoms, which can further complicate the differential diagnosis. Comprehensive imaging techniques such as CT and magnetic resonance imaging are needed to assess the extent of the tumor and its involvement of adjacent structures.[2]

Histopathological and ultrastructural examination of the resected mass in our case revealed characteristic features of ASPS, including positive staining for TFE3 and cathepsin K, consistent with previous studies on ASPS. Prominent Golgi apparatus, presence of rhomboid, or needle-shaped crystals with occasional neurosecretory

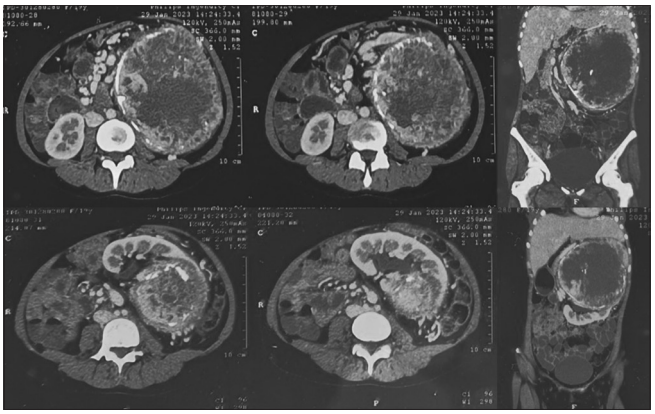

Figure 1: Axial and coronal sections of a heterogeneously enhancing left renal tumor

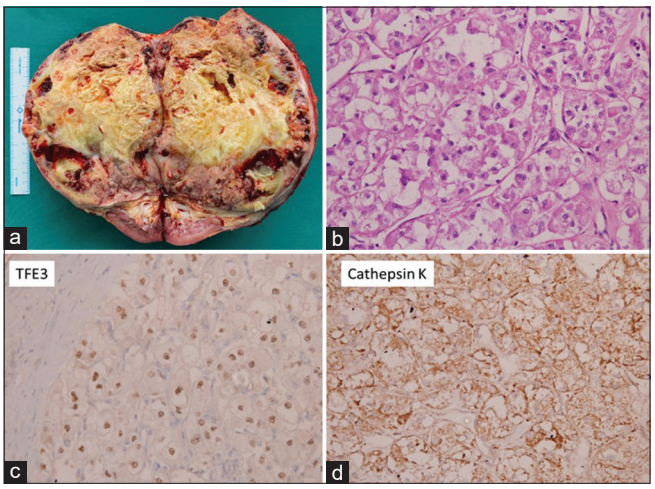

Figure 2: (a) Resected specimen, (b) Photomicrographs showing alveolar pattern with an abundant amount of eosinophilic granular cytoplasm, (c) Immunopositivity for TFE3 (nuclear), and (d) Cathepsin K (cytoplasmic)

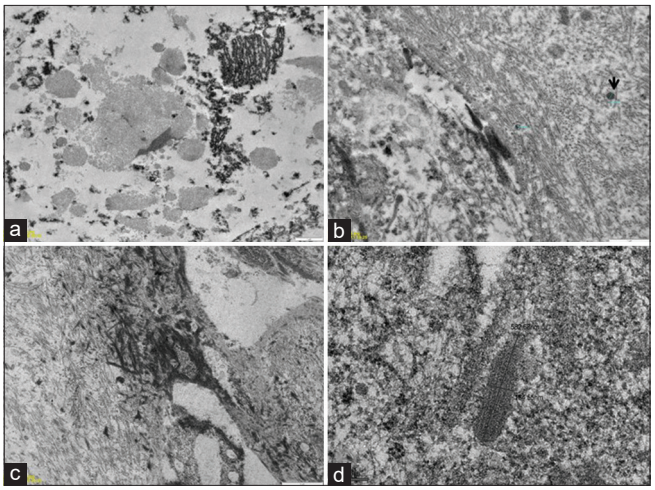

Figure 3: Electron micrographs: (a) Prominent Golgi apparatus, (b) Neurosecretory granules (arrow), (c) Numerous needle-shaped crystals, and (d) Rhomboid crystals

granules are characteristic features on electron microscopic examination.[3] These findings highlight the distinct immunophenotypic profile of ASPS and its differentiation from other renal malignancies.

ASPS is known for its propensity for metastasis, with rates reported in the literature from 43% to 72%. In our case, postoperative evaluation with PET-CT revealed metastasis to the left lung, along with recurrence in the left renal bed. These findings underscore the aggressive nature of ASPS and the importance of close surveillance and early detection of metastatic disease. The management of metastatic ASPS remains challenging, with limited treatment options available. Sunitinib, a multitargeted tyrosine kinase inhibitor (TKI), has demonstrated clinical benefit in patients with advanced, unresectable ASPS. It has shown objective response rates and disease stabilization in a significant proportion of patients. Other TKIs, such as pazopanib and cediranib, have also shown activity in ASPS and may be considered in certain cases.[4]

The management of ASPS is evolving, and ongoing research aims to identify additional therapeutic targets. Immune checkpoint inhibitors, such as pembrolizumab and axitinib, have shown promising results in some cases of ASPS. In addition, targeted therapies, such as directed at specific genetic alterations observed in ASPS, such as the ASPSCR1-TFE3 fusion gene, are being explored. The Food and Drug Administration (United States of America) has recently approved atezolizumab for adult and pediatric patients 2 years of age and older with unresectable or metastatic ASPS.[5]

In our case, the patient was initiated on sunitinib. However, the overall prognosis for metastatic ASPS remains guarded, necessitating ongoing research and the development of novel therapeutic approaches.

Due to the rarity of ASPS of the kidney, there is a paucity of data regarding its optimal management and long-term outcomes. The presented case contributes to the existing literature by providing insights into the clinical presentation, diagnostic challenges, and treatment strategies for this rare entity. The multidisciplinary approach involving surgical intervention, adjuvant therapy, and close follow-up is crucial in the management of ASPS. Furthermore, the identification of potential therapeutic targets and the exploration of novel treatment modalities are warranted to improve the prognosis of patients with ASPS of the kidney.

CONCLUSION

ASPS of the kidney is an uncommon malignancy with diagnostic challenges. These tumors tend to be aggressive with an increased propensity for metastasis. TKIs such as sunitinib, pazopanib, and regorafenib along with checkpoint inhibitors including pembrolizumab, nivolumab, and atezolizumab are emerging therapies in metastatic ASPS. Ongoing research is pivotal to improving outcomes for this rare renal neoplasm.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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