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Alveolar Soft Part Sarcoma Arising from the Kidney: Imaging and Clinical Features

Jung Myung Kim, MD[1], Soo Ah Im, MD[1], Soon Nam Oh, MD[1], Nak Gyun Chung, MD[2]

Departments of [1]Radiology and [2]Pediatrics, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul 137-701, Korea

Alveolar soft part sarcoma (ASPS) is an extremely rare malignant soft tissue sarcoma primarily affecting young patients. It usually occurs in the lower extremities, although it can occur in soft tissue anywhere in the body. However, to our knowledge, there has been no case of primary ASPS originating from the kidney in the literature. We herein present the imaging and clinical features of an ASPS which occurred in a 16-year-old male presented as a palpable mass in the left side of the abdomen.

Index terms: *Sarcoma; Alveolar soft part sarcoma; Kidney*

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a rare mesenchymal tumor, accounting for 0.5–1.0% of all soft tissue sarcomas (1). It mainly occurs in young adults aged between 15 and 35 years. This slowly growing, painless mass frequently accompanies metastatic lesions at the time of diagnosis. It usually affects the lower extremities in adults and the head and neck in children. It could occur in many other sites including the female genital tract, mediastinum, breast, urinary bladder, gastrointestinal tract, retroperitoneum and bones (2). Likewise, ASPS can arise in soft tissue anywhere in the body. However, no case of primary ASPS of the kidney has been reported yet. Here, we report a rare case of renal involvement of ASPS with a brief review of the literature.

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Corresponding author: Soo Ah Im, MD, Department of Radiology, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, 222 Banpo-daero, Seocho-gu, Seoul 137-701, Korea.
• Tel: (822) 2258-1455 • Fax: (822) 599-6771
• E-mail: saim@catholic.ac.kr

CASE REPORT

A 16-year-old male patient visited our hospital for the evaluation of left-sided abdominal pain and a palpable mass for three months. Ultrasonography (US) (iU22; Philips Medical Systems, Bothel, WA, USA) revealed a solid heterogeneously hyperechoic mass measuring more than 20 cm in the left mid-abdomen combined with internal increased blood flow and dilated vascular structures (Fig. 1A, B). The renal parenchyma maintained its normal structure in the upper aspect of the mass. Since the boundary between the mass and renal parenchyma was poorly defined, we assumed that the mass was arising from the left kidney. Enhanced abdominal computed tomography (CT) scans (Somatom Sensation 64; Siemens Medical Solutions, Erlangen, Germany) showed a huge enhancing mass (about 15 x 13 x 21 cm), involving almost the whole left kidney (Fig. 1C, D). The mass demonstrated a few faint calcifications and non-enhancing areas suggestive of necrosis. Although the upper portion of the kidney appeared to be normal even on US, the boundary of the mass seemed relatively indistinctive on CT. The left renal artery and a vein with variceal dilatation were observed around the mass and no distinctive filling defect was detected within the vessels. In addition to this mass in the left kidney, multiple

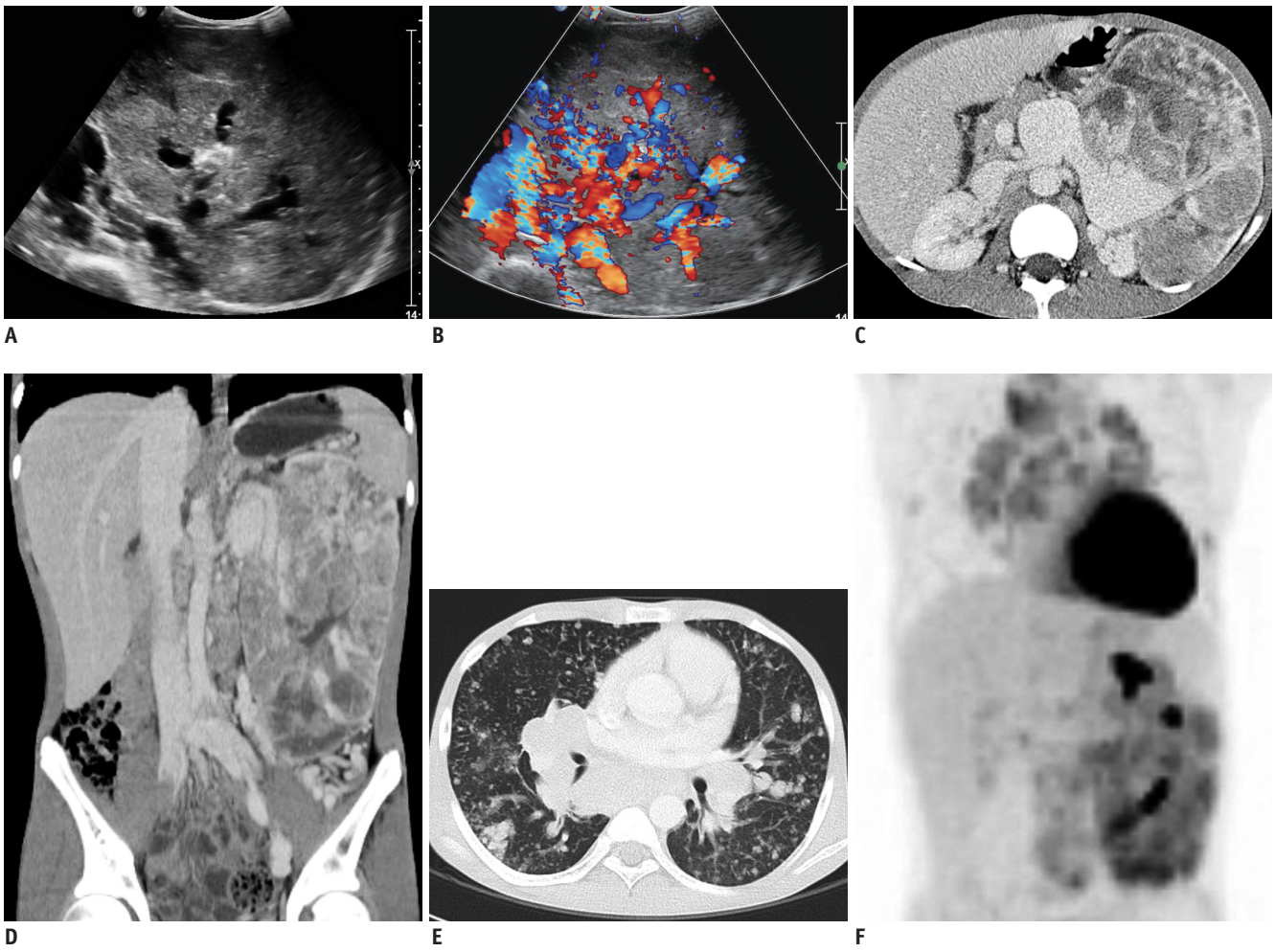

Fig. 1. Imaging and pathologic findings of alveolar soft part sarcoma arising from kidney in 16-year-old boy.
Two-dimensional **(A)** and Doppler **(B)** ultrasonographic images show heterogeneously hyperechoic mass in left kidney with internal and peritumoral dilated vascular structures. **C, D.** Contrast-enhanced CT scan of abdomen. Axial **(C)** and coronal **(D)** images show huge enhancing mass with internal necrotic foci, occupying almost whole left kidney. Numerous collateral vessels in addition to renal artery and vein show variceal changes. **E.** On chest CT with lung window setting, multiple different sized pulmonary nodules and diffuse interlobular septal thickening are noted in both lung fields, suggesting metastasis. **F.** PET-CT shows inhomogeneous FDG uptakes in large mass of left kidney (max SUV = 6.4) and in mass of small bowel mesentery (max SUV = 3.9). Multiple pulmonary nodules in both lungs and lymphadenopathies of left para-aortic, aortocaval and retrocrural regions, right supraclavicular region, mediastinum and in both perihilar regions showed FDG uptakes. FDG = fluorodeoxyglucose, max SUV = maximum standardized uptake value, PET = positron emission tomography

enlarged lymph nodes were found in the left retroperitoneal, left para-aortic, aortocaval and left retrocrural regions. At the same time, the chest radiography showed a mediastinal widening with contour bulging of both hila and diffuse nodular opacities in both lung fields. An enhanced chest CT (Somatom Sensation 64; Siemens Medical Solutions, Erlangen, Germany) revealed a mild interlobular septal thickening with randomly distributed multiple nodules in both lung fields. Moreover, extensive mediastinal and bilateral perihilar lymphadenopathies were detected (Fig. 1E). An osteolytic lesion was found in the glenoid cavity of the right scapula. According to the positron emission tomography (PET)/CT (Biograph 40 True Point; Siemens Medical Solutions, Knoxville, TN, USA) images acquired 60 minutes after injection of 8.3 mCi 18F-fluorodeoxyglucose (FDG), an inhomogeneous FDG uptake was detected in the large mass of the left kidney (maximum standardized uptake value [max SUV] = 6.4). Multiple pulmonary nodules in both lungs showed a max SUV of 1.5. The lymphadenopathies of the left retroperitoneal, left para-aortic, aortocaval and retrocrural regions, right supraclavicular region, mediastinum and both perihilar regions showed a max SUV from 2.8–3.9 (Fig. 1F).

An US-guided core needle biopsy was performed for the mass of the left kidney. The pathology findings of the biopsy specimen confirmed the diagnosis of ASPS; nests

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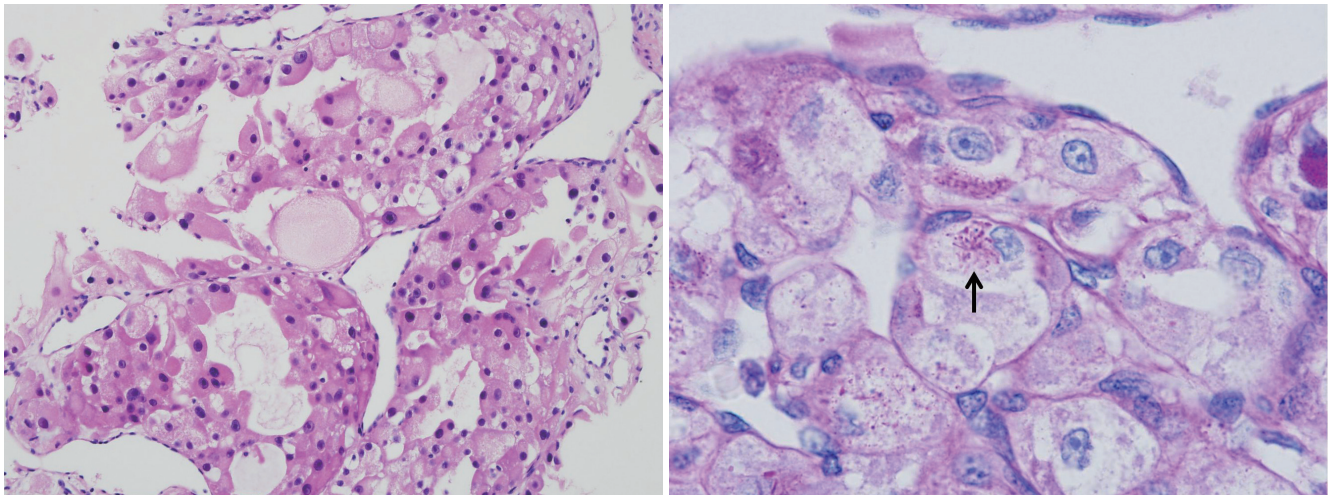

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Fig. 1. Imaging and pathologic findings of alveolar soft part sarcoma arising from kidney in 16-year-old boy.
G. Epithelioid tumor cells arranged in nests contain abundant eosinophilic cytoplasm (H&E, x 200). **H.** Tumor cells exhibit prominent periodic acid-Schiff (PAS)-positive, rod-shaped crystals (arrow) in cytoplasm (PAS Stain with diastase, x 1000).

of polygonal cells abundant with eosinophilic cytoplasm were separated by vascular structures (Fig. 1G), showing periodic acid-Schiff (PAS)-positive and diastase-resistant rod-shaped crystals within the cytoplasm (Fig. 1H). The suspicion of renal cell carcinoma (RCC) was ruled out as the specimen did not contain psammomatous calcifications on hematoxylin and eosin staining and the cells were negative for pancytokeratin and RCC antigen on immunohistochemical staining. Also no genetic abnormality was found in the additionally conducted cytogenetic study with karyotyping.

The patient received chemotherapy with ifosfamide and doxorubicin including multiple pulmonary lymph nodes and bone metastases after the diagnosis. The left kidney tumor showed no change on the enhanced CT which was performed for response evaluation after two chemotherapy sessions and the metastatic lymphadenopathies increased in size, exhibiting a further progression of the disease. The patient refused further treatment and died after two months.

DISCUSSION

Alveolar soft part sarcoma was first described by Christopherson et al. (3) in 1952. Histologically, round or polygonal cells abundant with eosinophilic cytoplasm are arranged in nests and each nest is separated by a coated vascular structure, exhibiting similar features to those of alveoli. The tumor cells are positive for PAS staining and rhombus- and rod-shaped crystals resistant to diastase are identified within the cytoplasm (2). Although a large number of studies have been conducted to find out the exact mechanism underlying the development of ASPS, the pathological mechanism still remains unclear. The name 'alveolar' soft part sarcoma is given based on the histological features of the tumors.

In general, ASPSs are negative for epithelial markers (i.e., cytokeratins and epithelial membrane antigen), specific neuroendocrine markers (i.e., chromogranin A and synaptophysin) and specific melanocytic markers (i.e., HMB45 and Melan-A). Non-specific markers such as neuron-specific enolase and vimentin may be positive in roughly 30–50% of the cases (2).

Alveolar soft part sarcoma is often characterized by its slow growing nature and tendency to metastasize. Approximately 20% of ASPSs accompany metastatic lesions at the initial diagnosis and approximately 80% of patients with ASPS experience metastasis during the periods of treatment and follow-up. The prevalence of metastasis is higher with a large tumor size at the initial diagnosis. ASPS tumors are larger in size and have a poor prognosis in adult patients (1). The size of the mass was 21 cm in our case and metastatic lesions were already present in the lungs, lymph nodes and bones at the time of diagnosis.

Extensive resection of the mass (R0 resection: complete resection with no microscopic residual tumor) is considered as the most effective treatment of ASPS. The local recurrence rate reportedly ranges from 10% to 30% after the removal of the mass and an adjunctive radiation treatment could be beneficial to prevent a relapse. The surgical resection of the lesion was considered to be difficult in our patient due to concurrent extensive

metastasis. For this reason, a conventional chemotherapy was initially attempted using cytotoxic agents. However, the effectiveness of conventional chemotherapy is known to be insignificant in ASPS according to several previous other studies (1). Some studies have reported the effectiveness of interferon alpha and recent studies have proved the effectiveness of anti-angiogenic agents (4).

Usually, ASPS could demonstrate a similar appearance as cavernous hemangioma or arteriovenous malformation, since there is an abundant distribution of blood vessels within the mass. ASPS can be differentiated from those similar-looking tumors as the progress of this malignant soft tissue tumor is totally different from others. The ASPS appeared as a solid mass with heterogeneous internal echogenicity and a rapid blood flow on US and exhibited a low resistance index due to the arteriovenous shunt within the internal mass. The CT scan showed a low or the same attenuation as the peripheral muscles on the pre-contrast scan and a dense enhancement was observed after contrast infusion. The signal intensity of the mass was slightly higher or the same as that of the surrounding muscles on T1-weighted MR images, and a high signal intensity of the lesion was seen on T2-weighted MR images. A dense enhancement was noted after the administration of contrast medium. The findings of a dilated feeding artery and particularly a draining vein around the masses strongly suggested the presence of ASPS. A flow void was seen inside and outside of the mass on T1- and T2-weighted MR images due to the prominent vascular structure (5). Our case also demonstrated the presence of numerous collateral vessels in the tumor and in the peritumoral region in addition to the renal artery and vein with variceal changes. The PET-CT facilitated the differentiation of the primary lesion from metastatic lesions in the presence of multiple masses at the time of initial diagnosis (6). In this case, the highest FDG uptake (max SUV = 6.4) was observed in the mass of the left kidney.

Considering the fact that the primary lesion involved the kidney and that the above mentioned radiological findings were manifested, ASPS needs to be differentiated from malignant renal tumors such as RCC, Wilms' tumor and other primary renal sarcomas including clear cell sarcoma and malignant rhabdoid tumor.

Renal cell carcinoma is an extremely rare childhood renal malignancy. However, it may be more frequently seen in the age between 15 and 19 years. Histologically, RCC may mimic ASPS by the virtue of their abundant eosinophilic to clear cytoplasm. An immunohistochemical evidence of strong cytokeratin and expression of RCC antigen makes it possible to distinguish RCC from ASPS (2). A large number of recent studies of molecular cytogenetics had confirmed the expression of ASPL-TFE3 fusion gene in ASPS. ASPL-TFE3 fusion gene is a fusion protein which is composed of ASPL gene on chromosome 17p25 and the TFE3 gene on chromosome Xp11. The expression of ASPL-TFE3 fusion gene and its type could play a crucial role in the discrimination between an ASPS occurrence in the kidney and a RCC variant. This gene is expressed as unbalanced der (17) t(X;17)(p11.2;q25) in ASPS while it is found as balanced t(X;17)(p11.2;q25) in some of RCC variants occurring in children or young adults (7). In this case, karyotyping was done with both tumor specimen and bone marrow, and no genetic abnormality was found.

Wilms' tumor, the most common pediatric renal malignancy, is hardly seen in adults aged over 16 years. The tumor commonly metastasizes into the lungs, liver and lymph nodes as in RCC. Furthermore, the tumor may extend to the renal vein and inferior vena cava (IVC). In our case, there was no evidence of tumor extension into the renal vein and IVC, although multiple metastases were found in the lungs, bones and lymph nodes (8).

Clear cell sarcoma and rhabdoid tumor are particularly uncommon pediatric neoplasms of the kidney. In the case of clear cell sarcoma, it mainly affects patients aged between 1–4 years, while rhabdoid tumor mostly occurs before the age of 2 years (9).

Unfortunately, radiological findings do not provide distinct characteristics for each of those tumors. Therefore, the pathological confirmation with a biopsy is crucial for making an accurate diagnosis. Like in other ASPSs, the findings of increased vascular flow and variceal dilatation of vessels within and around the mass, including the necrotic portion, were detected on a CT scan in our case. The US revealed the presence of a solid mass with heterogeneous echotexture, showing an increased vascular flow and variceal dilatation of vessels.

An ASPS is a very rare mesenchymal tumor and this is the first case report about an ASPS occurring in the kidney. Although it is an uncommon entity, ASPS needs to be included in the list of differential diagnosis when a renal mass displaying the above imaging findings is found in young adult patients.

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