Incidentally diagnosed retroperitoneal alveolar soft part sarcoma

A 20-year-old male incidentally diagnosed with left retroperitoneal mass on ultrasound. Computed tomography scan abdomen revealed heterogeneously enhancing left retroperitoneal mass with marginal blood vessels. Histopathological examination of postsurgical tumor specimen showed cells forming pseudoalveolar structures separated from each other by sinusoidal vascular channels suggestive of alveolar soft part sarcoma.

Key words: Computed tomography scan, pseudoalveolar pattern, retroperitoneal sarcoma

INTRODUCTION

Retroperitoneal sarcomas are rare tumors with common histological types in an adult being liposarcomas, leiomyosarcomas, and malignant fibrous histiocytomas (MFH).[1] Alveolar soft part sarcoma (ASPS) is an uncommon tumor of uncertain histogenesis, which occurs most often in the soft tissues of the pelvis and the lower limbs and is very rare in the retroperitoneal space.[2]

CASE REPORT

A 20-year-old young male presented with the pain left hypochondrium after hit by a cricket ball on his left lower chest. Ultrasound abdomen revealed a large heterogeneous hypoechoic mass lesion in left retroperitoneal cavity with moderate internal vascularity, inferomedially to the left kidney abutting the left psoas muscle. Rest intra-abdominal solid visceral organs were normal. Routine blood investigations were normal. Cross-sectional imaging with contrast-enhanced computed tomography (CECT) abdomen revealed a large (7 cm × 8 × 7 cm) well marginated heterogeneously enhancing left sided retroperitoneal mass lesion inferomedial to the left kidney [Figure 1]. Perilesional fat planes were well preserved with adjacent structures except with left psoas muscle. Solid visceral organs were normal on imaging. No significant lymph nodes were seen.

Intraoperatively, a retroperitoneal oval mass was seen just below the left kidney infiltrating the left psoas muscle. Tumor was excised with wide margins. On gross examination, capsulated mass with congested external surface was noted. On serial sectioning, [Figure 2] cut surface appeared variegated, lobulated with internal hemorrhagic cystic areas. Histopathology revealed large polygonal cells with eosinophilic cytoplasm, distinct cell borders with pleomorphic, and vesicular nuclei having prominent nucleoli. The cell nests were separated by fibrovascular septa giving a pseudoalveolar pattern. Hemorrhage and necrosis with tumor emboli in adjacent vessels were also seen. Immunohistochemistry showed focal cytoplasmic MYOD1 positivity and S-100 protein positivity and negativity for epithelial membrane antigen, vimentin, pan cytokeratin (excludes renal cell carcinoma (RCC), and chromogranin (excludes paraganglioma). Based on characteristic histopathological findings and immunohistochemical findings final diagnosis of ASPS was given. Postoperatively, patient received ifosfamide and doxorubicin-based adjuvant chemotherapy and radiotherapy. Follow-up CECT imaging of abdomen after 6 months, showed local recurrence of the mass with invasion of the adjacent left kidney. Multiple bony metastases were seen in lumbar and sacral vertebrae, right sixth rib and left upper femur shaft on skeletal survey. Unfortunately patient died within next 1-month.
DISCUSSION

Only 10–20% of sarcomas are retroperitoneal sarcomas. The most common types of retroperitoneal soft tissue sarcomas in adults are liposarcomas, leiomyosarcomas, and MFH.[1] ASPS is a rare tumor of uncertain histogenesis constituting 0.5–1.0% of all soft tissue sarcomas.[3] Christopherson et al. gave this term to the sarcoma, which had a characteristic pseudo alveolar pattern on histology formed by aggregates of large granular cells surrounded by vascular channels mimicking the alveolar pattern of respiratory alveoli.[4] It is hypervascular soft tissue tumor. It usually occurs in extremities in adult patients and head and neck region in children. Other unusual locations include the calf, upper extremity, retroperitoneum, female genital tract, and stomach.[1] The tumor affects primarily younger patients with a female preponderance; the peak age of incidence is between 15 and 35 years.[2]

Imaging findings are nonspecific. Non-contrast CT shows iso or hypodense mass compared to skeletal muscles. ASPS appears isointense or hyperintense compared to skeletal muscles on T1-weighted and heterogeneous hyperintense on a T2-weighted magnetic resonance imaging (MRI). ASPS shows strong enhancement on CECT and MRI (contrast-enhanced MRI [CEMRI]) with central necrosis in up to 75% of the cases. Increased perilesional and intrallesional vascularity can be seen in color Doppler ultrasonography, CECT, CEMRI, and/or in angiography.[3]

The differential diagnosis of lesions with high-flow vascular structures as a prominent feature and solid components by cross-sectional imaging is limited and include ASPS, rhabdomyosarcoma, extraskeletal Ewing sarcoma/peripheral neuroectodermal tumor, hemangioendothelioma, hemangiopericytoma, and synovial sarcoma.[9]

Majority of ASPS cases show classic histological patterns composed of uniform organoid nests of polygonal cells with focal pseudoalveolar arrangement. The tumor cells are rich in cytoplasm, containing periodic acid-Schiff and diastase-positive granules and typical crystals. The nuclei are round or oval, have an irregular chromatin pattern, and their nucleoli are clear, without frequent mitoses. Histologically RCCs, adrenal cortical carcinomas, and hepatocellular carcinomas may mimic ASPS.[9]

The histogenesis of the tumor remains uncertain due to variable immunohistochemistry findings. In general, ASPS remains negative for neuroendocrine, epithelial markers, and positive for myogenic markers (desmin/myoglobin/smooth muscle actin/vimentin/cyttoplasmic MyoD1).[5]

Complete surgical resection is the mainstay of therapy. The utility of adjuvant treatment with chemotherapy or radiotherapy remains uncertain. Postoperatively local recurrence as well as metastasis can occur.[9]

REFERENCES


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