Alveolar Soft Part Sarcoma Presenting as Hypervascular Adrenal Metastasis

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Abstract

Hypervascular adrenal masses include pheochromocytoma, metastases caused by clear renal cell carcinoma/hepatocellular carcinoma. Alveolar soft part sarcoma (ASPS) causing hypervascular metastases is not described in the literature. Here, we describe the first case of ASPS presenting as hypervascular metastasis. Our case was a 23-year-old male incidentally detected right adrenal mass during the evaluation of pain in the abdomen. On computed tomography (CT), adrenal mass showed bright enhancement in early arterial phase (unenhanced Hounsfield unit [HU]-45.3; arterial phase HU-158.2). 18- flurodeoxyglucose positron emission tomography/CT showed multiple lesions and was confirmed histologically to be due to ASPS.

Keywords: Adrenal contrast-enhanced computed tomography, hypervascular metastasis, soft tissue sarcoma, washout

Introduction

Hyper-vascular adrenal masses include pheochromocytoma, metastases caused by clear renal cell carcinoma/hepatocellular carcinoma. Alveolar soft part sarcoma causing hyper-vascular metastases is not described in the literature. Here we describe first case of alveolar soft part sarcoma presenting as hyper-vascular metastasis.

Case Report

A 23-year-old male patient presented with the early satiety, pain in the abdomen, and weight loss (5–6 kg) for the past 4 months. Ultrasonography (USG) abdomen showed large right suprarenal mass (9.5 cm × 8.8 cm × 12.5 cm) for which endocrinology referral was sought. The patient underwent multiphase contrast-enhanced computed tomography (CECT) which includes unenhanced, early arterial (20 s), early venous (60 s), and delayed (15 min) phases. CECT [Figure 1] showed rounded right suprarenal mass with central areas of necrosis and had a size of 9.9 cm (maximum size on axial images). Mass showed heterogeneous contrast enhancement, and it showed bright enhancement in arterial phase (Hounsfield unit [HU]-158.2) with good washout (Absolute washout percentage – 70.1%). On biochemical evaluation, plasma metanephrines (44.1 pg/ml, normal range <180 pg/ml) and normetanephrines (30.4 pg/ml, normal range <180 pg/ml) were normal ruling out secretory pheochromocytoma. I^{131} metaiodobenzylguanidine scan was done to rule out the possibility of nonsecretory pheochromocytoma, which showed no significant uptake in the mass. On morning 8 am cortisol (13.85 μg/dl, normal range <30 μg/dl) and overnight dexamethasone suppression cortisol (0.8 μg/dl, normal range <1.8 μg/dl) ruled out the secretory adrenal cortical carcinoma. 18-flurodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) revealed [Figure 2] multiple hypermetabolic lesions at right adrenal mass (maximum standardized uptake value [SUV_{max}]: 4.52), lytic lesion in the left

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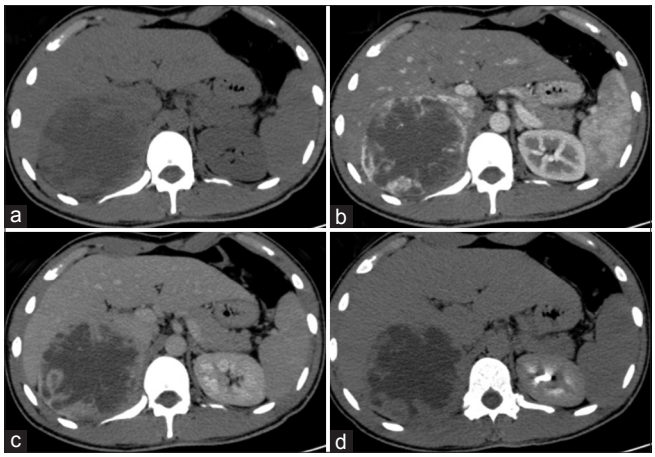

Figure 1: Multiphase CECT showing right adrenal mass with maximum diameter of 9.9 cm (a) unenhanced phase (HU of 45.3) with central areas of necrosis, (b) early arterial phase showing bright enhancement of mass (HU-158.2), (c) early venous phase (HU-98.6) showing washout as compared to early arterial phase, (d) 15-min delayed phase (HU-61.2) showing washout. CECT: contrast-enhanced computed tomography, HU: Hounsfield unit. Technique: 64-slice multidetector CT system (Brilliance 64, Philips Healthcare, Best, and the Netherlands) with imaging done at baseline, 20 s (early arterial), 1 min (early venous), and 15 min (delayed)

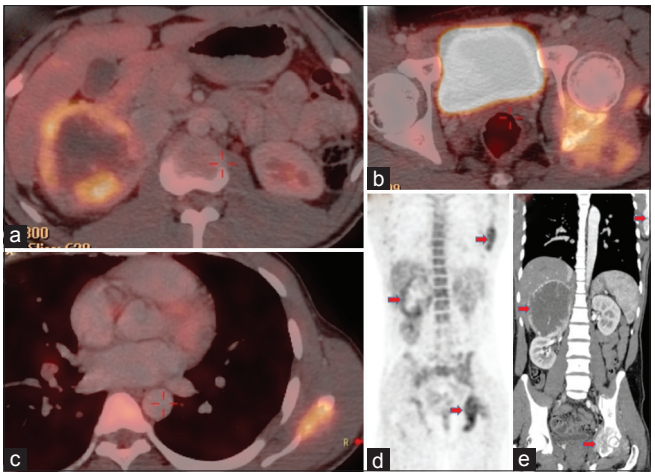

Figure 2: (a) ^{18}F-FDG avid lesion in the right adrenal mass (SUV_{max} 4.52), (b) ^{18}F-FDG avid lesion in the left gluteal region (SUV_{max}-5.5), (c) ^{18}F-FDG avid lesion in the left scapular region (SUV_{max} 4.6), (d and e) Coronal sections of FDG and CECT showing all three lesions (arrows), ^{18}F-FDG: 18-flurodeoxyglucose positron emission tomography/computed tomography, SUV_{max}: maximum standardized uptake value, CECT: contrast-enhanced computed tomography. Technique: whole-body 18-F-FDG PET/CT was performed in a BGO plus, full ring PET/CT scanner with intravascular contrast injection

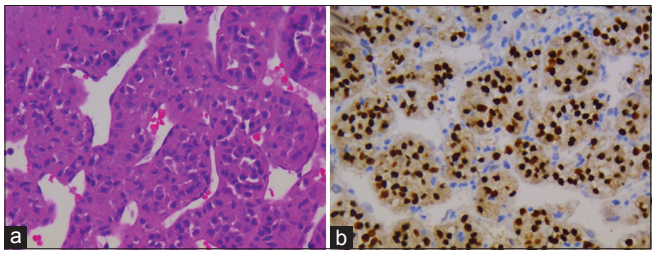

Figure 3: (a) Hematoxylin and eosin staining under light microscopy (×40) showing tumor cells arranged as nests separated by thin vascular septae with central round nucleus and moderate eosinophilic cytoplasm, (b) on immunohistochemistry, tumor cells showed positivity for TFE suggestive of alveolar soft part sarcoma

scapula (SUV_{max}: 4.46), and soft tissue mass (7.3 cm) noted in the left gluteal region eroding the left iliac bone, acetabulum, inferior pubic rami and possibly invading the hip joint (SUV_{max}-5.5). USG-guided biopsy from the adrenal and gluteal region was done. Both biopsy cores showed tumor cells arranged as nests separated by thin vascular septae. Cells were discohesive with central round nucleus and moderate eosinophilic cytoplasm on microscopy [Figure 3a]. On immunohistochemistry, tumor cells were positivity for TFE [Figure 3b] and epithelial membrane antigen (focal) and were negative for cytokeratin, synaptophysin, inhibin, melan-A, and PAX8 diagnostic of alveolar soft part sarcoma (ASPS). The final diagnosis was primary gluteal region ASPS with hypervascular adrenal metastasis. The patient was referred for palliative chemotherapy as the patient had distant metastases.

Discussion

In recent years, adrenal masses are receiving increased attention due to surge in the incidence of adrenal incidentaloma.[1] Classification of adrenal masses depending on the etiology has become important as management varies from observation to surgical resection.[2] Moreover, it is imperative to not miss a functional/malignant lesion, due to the low prevalence of such conditions.[2] Our patient had hypervascular adrenal lesion on computed tomography (unenhanced HU-45; arterial phase HU-158). 18F-FDG PET/CT showed multiple lesions and was confirmed histologically to be due to ASPS. CECT-derived absolute and relative washout percentages were used to differentiate adenoma and nonadenoma adrenal masses. However, the hypervascular adrenal masses usually do not follow these washout patterns.[3] Hypervascular adrenal masses constitute mainly pheochromocytoma and hypervascular metastasis. Pheochromocytoma is classically characterized as brightly enhancing adrenal masses, but it can have different characteristics on washout patterns from good washout to poor washout.[4] Hypervascular adrenal metastasis may have similar washout to adenomas on delayed contrast-enhanced CT images.[3] Clear cell renal carcinoma and hepatocellular carcinoma are described in the literature as hypervascular adrenal metastasis.[3] To the best of our knowledge, our case is first to report hypervascular adrenal metastasis caused by ASPS.

ASPS most often highly vascular and can present with metastasis, particularly in cases in the extremities.[5] In our case, both primary and adrenal metastasis showed similar high vascularity. This might be the reason for bright enhancement in the early arterial phase of CECT.

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Conflicts of interest
There are no conflicts of interest.

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