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Case Report

Primary adrenal leiomyosarcoma ☆

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ABSTRACT

Primary adrenal leiomyosarcoma is a very rare mesenchymal tumor that arises from smooth muscle cells in the wall of the central adrenal vein or its branches (1). Less than 50 cases have been published in the English literature (2). The tumors are aggressive and often metastasize. This report describes a case of primary adrenal leiomyosarcoma that presented as intermittent left flank pain of 6 months duration in an otherwise healthy 58-year-old Caucasian female. The patient was initially imaged with an abdominal ultrasound, which revealed a left suprarenal mass. A follow-up CT of the abdomen and pelvis confirmed a malignant appearing left adrenal mass. A subsequent PET-CT demonstrated increased metabolic activity within the adrenal mass without evidence of metastasis. Biopsy proven metastasis eventually was detected on surveillance CT studies over the course of 2.5 years. Since this is such a rare malignancy, documenting its imaging findings with multiple modalities is of importance to add to the medical literature and help further characterize its imaging features.

Introduction

Primary adrenal leiomyosarcoma is an extremely rare mesenchymal tumor that arises from smooth muscle cells that reside in the wall of the central adrenal vein or its branches [1]. A recent review of case reports found less than 50 cases have been reported in the English medical literature [2]. Risk factors include acquired immunodeficiency syndrome (AIDS) and Epstein-Barr virus infection [2]. The most common symptoms are abdominal/flank pain and anorexia and/or weight loss [2].Typical imaging findings are a heterogeneous adrenal mass with irregular margins that may have areas of necrosis. There may also be venous invasion and infiltration of adjacent organs [2]. The tumors are FDG avid on PET-CT imaging indicating increased metabolic activity.

Case report

A previously healthy 58-year-old Caucasian female with no significant past medical history complained of intermittent left flank of 6 months duration. She had no other complaints or laboratory abnormalities. She was initially evaluated with an abdominal ultrasound (Fig. 1). This revealed a lobulated left suprarenal mass.

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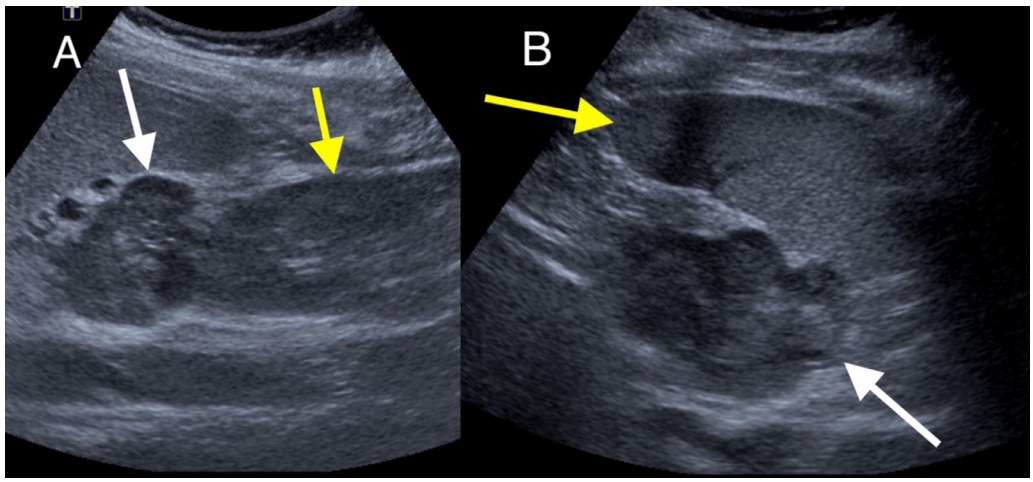

Fig. 1 – Abdominal ultrasound. (A) Longitudinal view: lobulated suprarenal mass (white arrow) isoechoic with the adjacent left kidney (yellow arrow). (B) Transverse view: lobulated suprarenal mass (white arrow) medial to the spleen (yellow arrow).

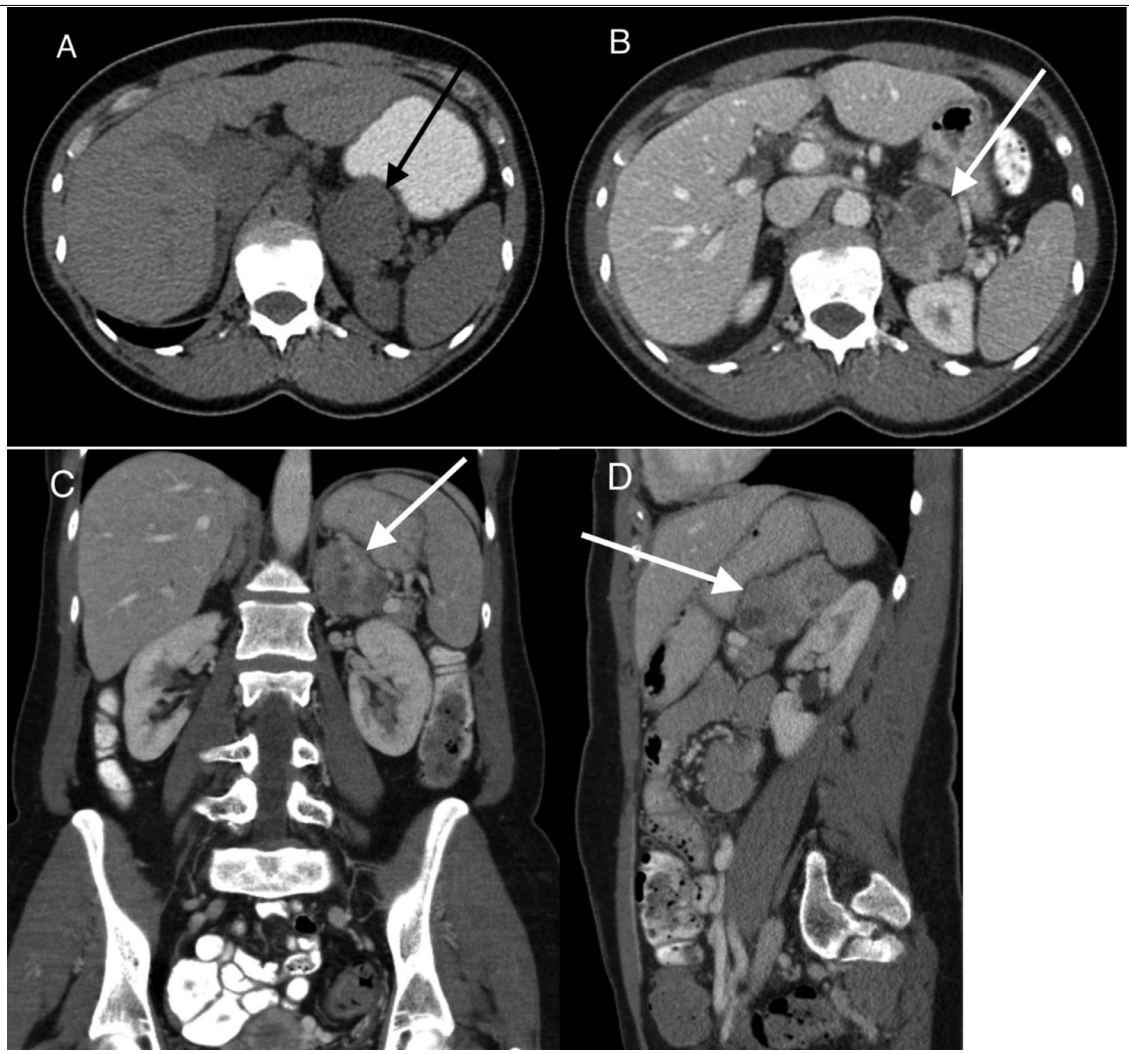

Fig. 2 – CT abdomen and pelvis: (A) Nonenhanced axial image: Lobulated suprarenal mass adjacent to the spleen (black arrow). (B) Contrast-enhanced axial: lobulated enhancing heterogeneous solid and cystic left adrenal mass (white arrow). (C) Contrast-enhanced coronal image: heterogeneous lobulated left adrenal mass (white arrow). (D) Contrast-enhanced sagittal image: heterogeneous lobulated mass with cystic areas indicating focal areas of necrosis (white arrow).

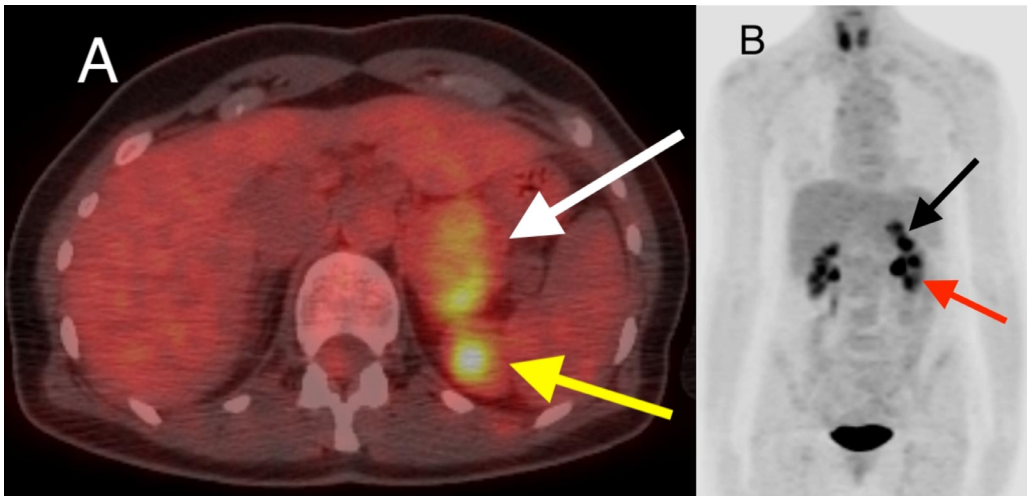

Fig. 3 – (A) FDG PET/CT axial fused image: hypermetabolic left adrenal mass (white arrow), left kidney (yellow arrow). (B) Coronal MIP PET image: hypermetabolic left adrenal mass (black arrow), left kidney (red arrow).

Table 1 – Tumor marker status.

Marker	Result
Smooth muscle actin	Most cells positive
Desmin	Positive
Calponin	Positive
Vimentin	Focally positive
Caldesmon	Positive
S100	Negative
CD34	Negative
ckit	Negative
Ki-67	Increased

This prompted further evaluation with a CT of the abdomen and pelvis (Fig. 2). The CT examination demonstrated a malignant appearing, enhancing, heterogeneous solid and cystic, lobulated 5.5 cm × 4.4 cm left adrenal mass. No calcifications or venous invasion were present. There was no evidence of abdominal or pelvic metastasis.

A preoperative FDG PET/CT was then performed (Fig. 3). The left adrenal mass exhibited increased metabolic activity resulting in a maximum SUV of 5.9 g/mL. There was no evidence of local or distant metastasis.

The mass was resected laparoscopically. The gross pathological specimen revealed a 7.5 cm × 4.0 cm × 4.0 cm, 56 g, well encapsulated adrenal mass with areas of necrosis. Margins were negative. No lymph nodes were involved. Histologic features were consistent with a leiomyosarcoma. Tumor markers are listed in Table 1.

Discussion

Primary adrenal leiomyosarcoma is an extremely rare mesenchymal tumor that arises from smooth muscle cells located in the wall of the central adrenal vein or its branches [1]. It was first reported in 1981 [3]. Since then, less than 50 cases have appeared in the medical literature [1]. Unfortunately, the tumor is usually not detected until an advanced stage because it is not hormonally active and there are no specific tumor markers [4]. Acquired immunodeficiency syndrome and Epstein-Barr infection may be predisposing conditions [2,5]. The median age at diagnosis is 60 years. There is no sex or side predilection [2]. Metastasis usually occurs to the lungs, liver, pancreas and bones [2]. Surgical resection is the standard of care [2,6]. Chemotherapy and radiation therapy are of uncertain benefit [1].

There is limited information describing the imaging characteristics of primary adrenal leiomyosarcoma because it is such a rare tumor and it has been reported only relatively recently. Therefore, this case report is of clinical significance because it adds to the growing body of medical literature by describing the imaging findings of primary adrenal leiomyosarcoma with multiple imaging modalities.

Patient consent

Written consent was obtained directly from the patient.

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