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Angiomyolipoma of the Adrenal Gland: A Case Presentation and a Review of Adrenal Lipomatous Tumors

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ABSTRACT

Angiomyolipoma (AML) is a typically benign renal tumor derived from mesenchymal tissue. Extrarenal occurrences of AML are possible, but the adrenals are an exceedingly rare site. To date, a total of 4 cases of adrenal AML have been documented in the English literature.[1–3] We present a case of right-sided adrenal AML found in a patient who initially presented with right-sided flank pain. Differential diagnosis of adrenal masses should include lipomatous tumors, as operative considerations and prognoses can be drastically altered.

Introduction

Angiomyolipoma (AML) is a typically benign renal tumor derived from mesenchymal tissue. As the name suggests, these tumors usually consist of 3 components: blood vessels, muscle cells, and fat tissue.[4] Extrarenal occurrences of AML are possible, and are most commonly found to occur in the liver.[1] Other possible extrarenal sites include the spleen, lungs, retroperitoneum, bone, and ovaries.[5] The adrenals are an exceedingly rare site of AML. To date, a total of 4 cases of adrenal AML have been documented in the English literature.[1–3] We present a case of right-sided adrenal AML found in a patient who initially presented with right-sided flank pain.

Case report

The patient is a 48-year-old woman who presented with right-sided flank pain. Outside evaluation revealed a 7-inch right-sided mass enveloping the superior portion of the kidney without IVC or renal vein involvement on CT imaging seen in Fig. 1. The differential included adrenal myelolipoma, adrenal cortical carcinoma, functional adrenal tumor, adrenal liposarcoma, and angiomyolipoma of the right kidney. She had previously undergone unsuccessful IR embolization of the mass due to suspicion of renal AML. Biopsy was deferred due to the patient's status as a Jehovah's Witness and the high-risk of encountering significant bleeding during the procedure.

A full adrenal workup was performed, including urine and plasma metanephrines, cortisol, aldosterone, and renin. All values were found to be within normal limits and the patient proceeded with a right-sided adrenalectomy. The mass, seen in Fig. 2, was able to be removed with minimal intraoperative complications. Her

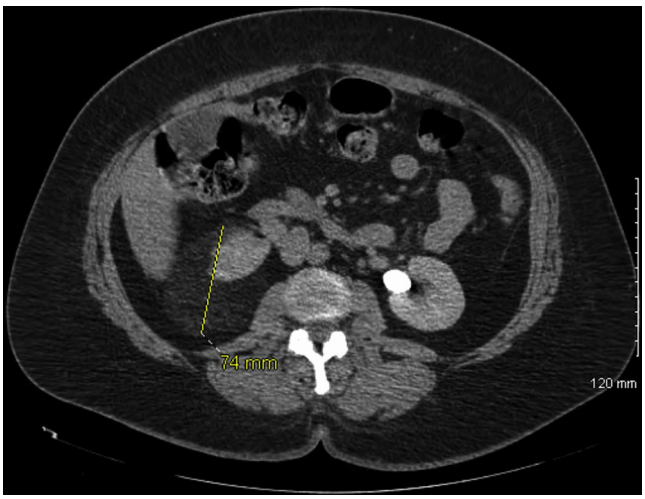

Figure 1. CT imaging, revealing a roughly 7 cm mass enveloping the right kidney without apparent renal vein or IVC involvement.

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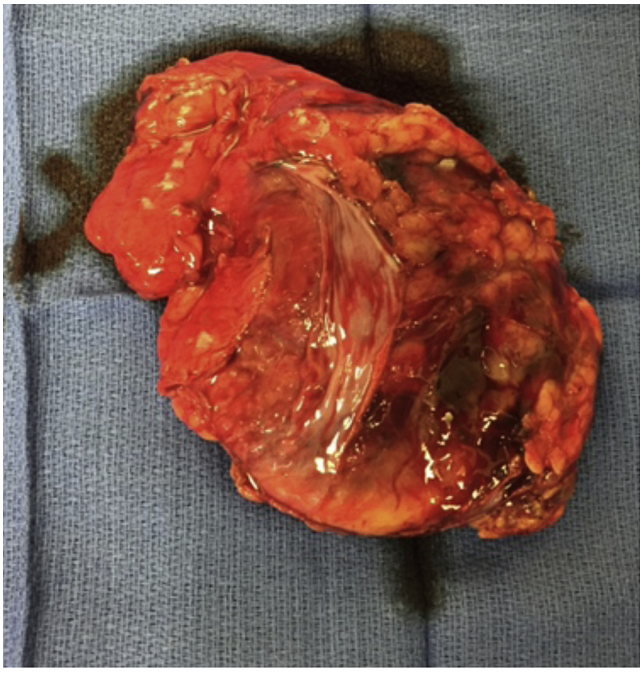

Figure 2. Gross image of the 10 cm resected mass.

postoperative course was generally unremarkable and she was discharged home on postoperative day 4.

Gross exam revealed a 10 cm, 290 g adrenal mass containing lipomatous elements without malignant changes. Microscopic evaluation, seen in Fig. 3 confirmed the presence of lipomatous tissue and also found areas of fat necrosis, some areas of focal intravascular emboli, and no hematopoietic elements suggestive of angiomyelolipoma. Samples were sent for MART-1 antibody staining due to the concern for possible adrenal AML, which returned positive. Due to the rarity of liposarcomatous tumors of the adrenal gland, samples were sent for external review. External review identified clusters of spindle cells in addition to thick walled blood vessels. Staining for HMB-45 and Desmin were also performed, confirming the diagnosis of AML.

Discussion

AML is an uncommon tumor, with an incidence of 0.13% in the general population and twice the incidence in females compared to males.[4] Although AML may arise spontaneously, there is an association with lymphangioleiomyomatosis (LAM) and tuberous sclerosis (TS). Up to 90% of AML cases occur in patients with a history of TSC. However, there have been no studies establishing a clear link between extrarenal occurrences of AML and a diagnosis of TSC.

Renal AML is included on a spectrum of mesenchymal tumors derived from perivascular epithelioid cells (PEC). The three components that compose AML are dysmorphic blood vessels epitheliod smooth muscle cells, and mature adipose cells. Diagnosis of AML can be made histologically using these features in addition to immunohistochemical stains with HMB-45 and MART-1/Melan-A antibodies. Usually, however, the diagnosis is made with imaging. The most easily identifiable characteristic on CT is macroscopically detectable amounts of hypodense fat.[4] On MRI, the typical finding is bright signal intensity on non–fat-suppressed images and dropout of signal with fat suppression.

AML typically presents with back/flank pain and hematuria. The more drastic presentation includes retroperitoneal hemorrhage from the tumor due to the friable blood vessels. The goal of treatment in such a case is to achieve hemostasis and prevent further complications. Options for treatment when the tumor is symptomatic include tumor excision, angioembolization, or cryotherapy. Active surveillance with serial ultrasounds is typically sufficient in patients with small lesions or who are asymptomatic.

Lipomatous tumors of the adrenal gland account for roughly 5% of primary adrenal tumors.[2] This group includes myelolipomas, lipomas, teratomas, angiomyolipomas, and liposarcomas. They usually present as either incidental findings on imaging or clinically with flank/back pain. The main concern with these tumors is spontaneous rupture leading to hemorrhage. In addition, epithelioid AML and liposarcoma are both malignant and aggressive lesions that are important to diagnose. Although AML is usually benign, it is important to maintain oncologic control intraoperatively due to the possibility of such malignant diagnoses.

This case represents the fifth case of adrenal AML in the English literature.[1–3] An additional six cases have been found in foreign publications. These cases are described in Table 1 below. Of note, there is a considerable predilection for female gender and age 40–50 year-old. These masses tend to be large (>10 cm). Our presented case is unique in that it arose sporadically without any other documented history or associated conditions. It also presented diagnostic challenges preoperatively and therapeutic challenges intraoperatively due to the patient's refusal of blood transfusions due to her status as a Jehovah's Witness.

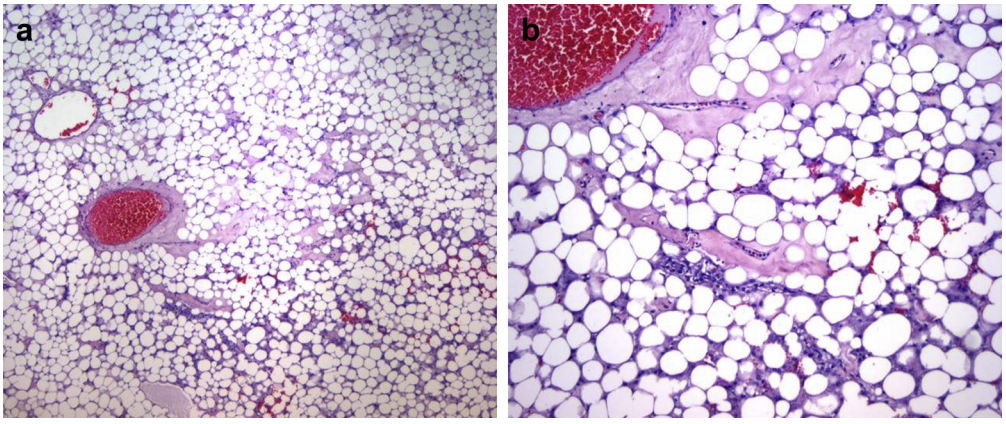

Figure 3. a: 4× magnification of H&E stained slide. Note the abundant lipomatous elements and the thick walled blood vessel. b: 10× magnification of H&E stained slide from previous image. Closer view reveals minor amounts of spindle cells interspersed between lipomatous elements.

Table 1
Compilation of previously published case reports of adrenal AML, starting with those published in English language journals, followed by foreign publications

Country published	Author/year	Age/sex	Size/weight	Presentation	Remarks	Follow-up
China	Lam, 2001[2]	46 yo, F	8 cm/115 g	Incidental on CT	-	12 mos
China	Lam, 2001[2]	20 yo, M	0.2 cm/3 g	Incidental on nephrectomy	TSC with multiple, bilateral renal AML's and hepatic AML	96 mos
USA	Elsayes, 2005[1]	49 yo, F	12 cm/-	Incidental on CT	TSC with small contralateral renal AML	-
USA	Kwazneski, 2016[3]	65 yo, F	13 cm/626 g	RUQ Abd pain	-	36 mos
USA	Antar, 2016	48 yo, F	10 cm/290 g	Flank pain	-	7 mos
Switzerland	Sutter, 2006[6]	32 yo, F	-/-	Abdominal pain	LAM with bilateral renal AML's	-
India	Godara, 2007[7]	45 yo, F	15 cm/-	Epigastric pain	-	18 mos
Italy	D'Antonio, 2009[5]	42 yo, M	6 cm/100 g	Flank, back pain	Monotypic epithelioid AML	12 mos
Turkey	Yener, 2011[8]	45 yo, F	5 cm/-	Epigastric pain	-	3 mos
England	D'Amico, 2012[9]	47 yo, F	10 cm/-	Incidental on ultrasound	Also with uterine leiomyoma	-
India	Sharma, 2014[10]	43 yo, F	10 cm/-	Groin pain	-	-

Conclusion

This case presents an exceedingly rare diagnosis of extrarenal angiomyolipoma of the right adrenal gland, representing only the fifth documented case to be described in the English literature. It underlines the importance of maintaining a large differential diagnosis and a high index of suspicion for possible malignant diagnoses. In this particular case, we also highlight hematologic considerations in a high-risk patient population. These tumors are typically managed with adrenalectomy or complete resection of the tumor due to presenting symptoms or suspicion of more insidious diagnoses. As more cases are described, it is likely that radiologic, pathologic, and surgical considerations will continue to be addressed.

Conflicts of Interest

None.

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