CASE REPORT

10.5005/jp-journals-10002-1040

Symptomatic Giant Adrenal Myelolipoma

Blake B Anderson, Lance J Hampton, Corey M Johnson, Georgi E Guruli

Division of Urology, Virginia Commonwealth University School of Medicine, Richmond, Virginia, USA

Correspondence: Georgi E Guruli, VCU, Division of Urology, 1200 E, Broad Street, West Hospital, 7th Floor, PO Box 980118 Richmond, VA 23298-0118, USA, Phone: (804) 828-5320, e-mail: gguruli@cedar.vcu.edu

ABSTRACT

In this article, we review the management of giant adrenal myelolipoma and review the literature in regard to incidence and management. Patient presented with abdominal fullness and discomfort with CT revealing a large, well-circumscribed right-sided suprarenal mass. A thoracoabdominal incision allowed exploration and removal of the mass together with the right adrenal gland. Pathology revealed the mass to be a giant adrenal myelolipoma, a benign but rare tumor.

Keywords: Adrenal myelolipoma.

INTRODUCTION

Adrenal masses are not uncommon, being reported in 5% of patients.[1] Often they are found as asymptomatic "incidentalomas" when patients undergo different types of imaging for a nonadrenal indication.[2] Less commonly, patients are symptomatic, as in the present case, due to the effects of hormone excess in functional tumors or due to mechanical compression as a result of tumor size. In the case presented here, the tumor was found to be a large unilateral primary adrenal ML, measuring 23.8 cm in its largest diameter on final pathologic examination. Adrenal ML is a rare, benign, and almost always nonfunctioning tumor comprised of mature adipose and hematopoietic structures.[3] To our knowledge, there are only 11 cases of giant adrenal ML reported in the literature.[3,4]

CASE REPORT

A 35-year-old man with a history of hypertension presented to our urology clinic with vague, right-sided abdominal discomfort that had been progressively worsening for 6 months. Patient was referred after ultrasound revealed a large solid hyperechoic right suprarenal mass. He underwent an abdominal CT scan (Fig. 1), which demonstrated a well-circumscribed, mixed fat and soft tissue lesion (-85 HU) arising from the right adrenal gland that measured 14.5 cm anteroposterior (AP) × 15.5 cm transverse × 17.4 cm craniocaudal. Also on CT, the right kidney was found to be shifted inferomedially by the tumor but otherwise looked normal. Adrenal ML was the most likely diagnosis. Chest imaging was within normal limits. A complete metabolic panel, complete blood count with differential, serum cortisol, urine aldosterone, urine metanephrine, and urine normetaphrine were all within normal limits. Unilateral adrenalectomy was offered due to tumor size as well as the possibility for further growth leading to spontaneous rupture and hemorrhage. Also, the inability to rule out malignancy (i.e. liposarcoma) necessitated surgical intervention.

Due to the large size of the mass, the patient was explored with an open approach via a thoracoabdominal incision with the patient in the full flank position. The incision began in the posterior axillary line in the intercostal space between the 10th and 11th ribs and came approximately 4 cm past the costal margin of the thoracic cage. The retroperitoneal space was entered allowing visualization of Gerota's fascia, which was then opened exposing the mass. Using blunt dissection, the well-circumscribed mass was mobilized. Significant tethering was identified around the medial portion of the mass. The right kidney was identified, being well-perfused albeit displaced inferomedially by the mass. The mass was then removed *en bloc* with the right adrenal gland. The right adrenal vein was suture ligated with 5-0 Prolene. A running 4-0 PDS suture was used to reapproximate the pleura and diaphragm while a 6-French red rubber catheter was used to evacuate the pneumothorax. A chest tube was not placed. A 1-0 Maxon suture was used to bring together the 10th and 11th ribs, and then the patient was closed in the normal fashion. The peritoneal space was never entered.

The *en bloc* specimen (Fig. 2) weighed 1575 gm with the adrenal mass measuring 23.8 × 11.6 × 7.5 cm and being variably

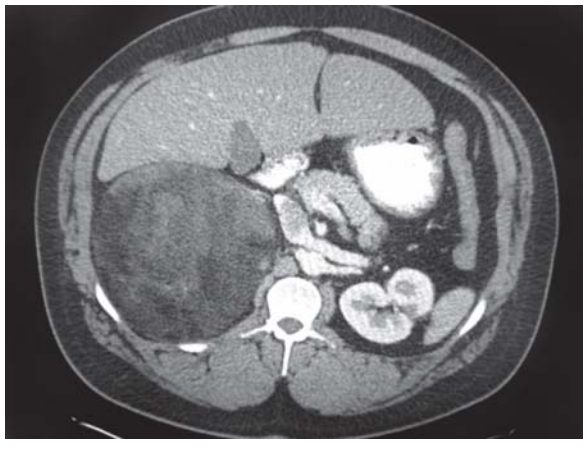

Fig. 1: Giant right adrenal mass

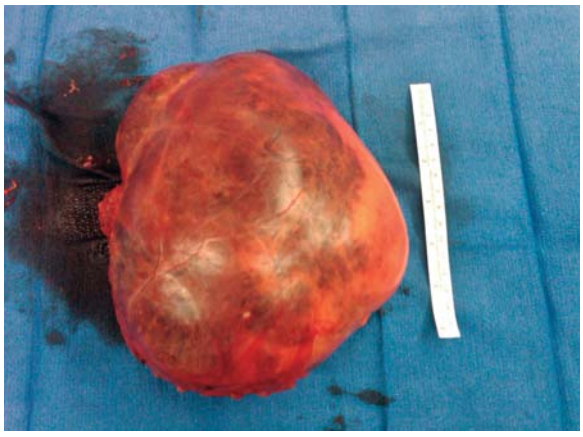

Fig. 2: Right adrenalectomy specimen

soft and firm, well-circumscribed, heterogeneous, and made up of yellow-tan and red-brown tissue. Its origin was determined to be the adrenal cortex. The adrenal gland was otherwise found to be uninvolved and compressed measuring 4.2 × 2.3 × 0.2 cm. Microscopic examination confirmed the diagnosis to be adrenal ML.

Patient required no blood transfusions and was immediately extubated following surgery. Postoperative chest X-ray showed no pneumothorax. Hospital course was uneventful with discharge on postoperative day 4. The patient has experienced no complications and no chronic pain.

DISCUSSION

As mentioned previously, adrenal ML is a rare tumor. Its prevalence upon autopsy has been reported as 0.08 to 0.2%, but it also constitutes 7 to 15% of adrenal incidentalomas.[2] With modern imaging modalities like CT, it seems that these tumors are less rare than once thought.[2] A recent retrospective review of CT reports on 65,231 patients revealed an adrenal mass prevalence of 5% with adrenal ML (6%) being the second most common type of mass behind adrenal adenoma (75%).[1]

Adrenal ML is most often diagnosed in patients in the 4th to 7th decades,[3,4] but this type of tumor has been identified in a wide range of patients (12 to 93 years old).[3] There is no clear gender predilection for adrenal ML in the existing literature[3]. The size of these tumors can vary significantly from microscopic diameters up to 40 cm in diameter.[5] While the location for ML is most often adrenal, they have been found in extra-adrenal locations, most often retroperitoneal.[3] Concerning laterality, some have observed that adrenal ML is more often right-sided with one study reporting 53 of 72 tumors as right-sided.[6]

Importantly, the pathophysiology of adrenal ML remains unclear. The prevailing theory is adrenocortical metaplasia of reticuloendothelial cells after exposure to different stimuli, including stress, inflammation, infection, and necrosis.[3] Some have investigated associations of adrenal ML with medical conditions, such as diabetes, hypertension, obesity, and malignancy, which could fit into the aforementioned theory as different causes of chronic adrenocorticotropic hormone (ACTH) release.[3] A review of the literature by Umpierrez et al suggests that patients, who have dysfunction of the pituitary-adrenal axis, such as those with Cushing's syndrome or 21-hydroxylase deficiency, may have greater risk of developing adrenal ML.[7]

Interestingly, adrenal ML is usually asymptomatic. One case series by Han et al demonstrated 15 out of 20 patients with adrenal ML to be asymptomatic.[3] They also found no association between symptoms and tumor size. This observation gains support from Mukherjee et al, who described the largest asymptomatic adrenal ML yet to be reported weighing 5.5 kg and measuring 28 × 26 × 17 cm.[4] This discrepancy between symptoms and size adds doubt to mechanical compression as a cause of pain, but individual patient and tumor characteristics must be taken into account. For example, in contrast to Han et al, Del Gaudio and Solidoro,[8] and Dieckmann et al[9] did report a correlation between tumor size and symptoms. Others suggested the causes of pain with adrenal ML are retroperitoneal hemorrhage, intratumoral hemorrhage, and tumor necrosis. Despite several documented reports of hemorrhage, there is no clear association between tumor size and risk for rupture.[3]

Although adrenal ML has been considered nonfunctional in the past, there has been at least one reported case of functional adrenal ML by Tamidari et al,[10] in which a tumor presenting symptomatically as a pheochromocytoma with 24-hour urine metanephrines of 2.86 mg/day was found by histology to be myelolipoma. This supports the use of a preoperative endocrine workup. No set guidelines exist for management of adrenal ML, but most authors agree that asymptomatic tumors smaller than 5 cm can be followed radiologically, whereas large or symptomatic tumors warrant surgical removal in keeping with individual treatment considerations.[3,4]

REFERENCES

1. Song JH, Chaudhry FS, Mayo-Smith WW. The incidental adrenal mass on CT: Prevalence of adrenal disease in 1,049 consecutive adrenal masses in patients with no known malignancy. AJR. 2008;190: 1163-68.
2. Kloos RT, Gross MD, Francis IR, Korobkin M, Shapiro B. Incidentally discovered adrenal masses. Endocr Rev 1995;16:460-84.
3. Han M, Burnett AL, Fishman EK, Marshall FF. The natural history and treatment of adrenal myelolipoma. J Urol 1997;157:1213-16.
4. Mukherjee S, Pericleous S, Hutchins RR, Freeman PS. Asymptomatic giant adrenal myelolipoma. Urol J 2010;7:66-68.
5. Polamaung W, Wisedopas N, Vasinanukorn P, Pak-art P, Snabboon T. Asymptomatic bilateral giant adrenal myelolipomas: Case report and review of literature. Endocr Pract 2007;13:667-71.
6. Kenney PJ, Wagner BJ, Rao P, Heffess CS. Myelolipoma: CT and pathologic features. Radiology 1998;208:87-95.
7. Umpierrez MB, Fackler S, Umpierrez GE, Rubin J. Adrenal myelolipoma associated with endocrine dysfunction: Review of the literature. Am J Med Sci 1997;314:338-41.
8. Del Gaudio A, Solidoro G. Myelolipoma of the adrenal gland: Report of two cases with a review of the literature. Surgery 1986;99:293-301.
9. Dieckmann KP, Hamm BP, Jonas D, Bauer HW. Adrenal myelolipoma: Clinical, radiologic, and histologic features. Urology 1987;29:1-8.
10. Tamidari H, Mishra AK, Gupta S, Agarwal A. Catecholamine secreting adrenal myelolipoma. Indian J Med Sci 2006;60:331-33.