Elmer Press

Case Report

J Endocrinol Metab. 2015;5(5):304-308

Adrenal Leiomyoma Mimicking Adrenal Malignancy: Diagnostic Challenges and Review of Literature

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Abstract

Leiomyoma arising from adrenals are extremely rare with less than 20 cases reported in medical literature. Leiomyomas, though benign lesions, may frequently be confused with a malignancy, both clinically and on imaging studies, especially when they occur at atypical sites like the adrenals. We present a 42-year-old lady with cachetic symptoms, abdominal swelling and dragging sensation for 6 months, having a 10 cm diameter non-tender firm palpable abdominal mass, which on computed tomography was diagnosed to be a 12.2 × 10.3 × 8.0 cm heterogeneously enhancing adrenal mass having absolute and relative contrast washout values suggestive of malignancy. The tumor was hormonally inactive. Left adrenalectomy was done. Resected specimen weighed 91 g measuring 12 × 10 × 8.0 cm. Histopathology revealed a well-circumscribed and encapsulated benign spindle cell arranged in fascicles and whorls confirming leiomyoma. In the last evaluation 6 months after surgery, there was no evidence of tumor recurrence. This is the largest adrenal leiomyoma ever reported. Leiomyomas have a varied age of presentation (2 - 72 years, median: 38) with female preponderance. They are usually unilateral and hormonally inactive. Human immunodeficiency virus and Epstein-Barr virus infections have been observed in 44.44% and 16.67% of cases respectively. However, direct link between immunodeficiency and adrenal leiomyoma has not been established. Adrenal leiomyoma can present as huge abdominal masses clinically mimicking malignancy, and radiologic investigations can also be misleading. Adrenal leiomyoma should always be considered in the differential diagnosis of huge unilateral non-functional incidentally detected adrenal lesions.

Keywords: Adrenal leiomyoma; Carcinoma; Malignancy; Neoplasm; Incidentaloma

Introduction

Leiomyomas are benign tumors arising from smooth muscle cells lining various organs in the body, most commonly observed in uterus followed by the gastrointestinal tract [1, 2]. Leiomyoma arising from adrenals are extremely rare with less than 20 cases reported in medical literature, to the best of our knowledge [1-18]. Adrenal leiomyomas are clinically asymptomatic, hence they usually present as incidentalomas [3, 16, 17]. Adrenal incidentalomas have been documented in approximately 1% of all abdominal computed tomography (CT) scans [19-21]. Leiomyomas, though benign lesions, may frequently be confused with a malignant tumor, both clinically and on imaging studies, especially when they occur at atypical sites like the adrenals. This report presents a middle-aged lady with cachetic symptoms, presenting with a palpable huge abdominal mass, which was subsequently diagnosed to be adrenal leiomyoma. A subsequent review of all available literature on adrenal leiomyoma was also done.

References for review of literature on adrenal leiomyoma were identified through searches of PubMed, Medline, and Embase for articles published to September 2015 using the terms "adrenal leiomyoma" (MeSH Terms) OR "adrenal leiomyoma" (all fields). The reference lists of the articles thus identified were also searched. The search was not restricted to English-language literature.

Case Report

A 42-year-old lady presented with loss of appetite and weight (12 kg in 4 months) along with heaviness and dragging sensation on left side of her abdomen of 6-month duration. Examination was significant for body mass index of 17.2 kg/m^2, normal blood pressure and a visible abdominal swelling which on palpation revealed a 10 cm diameter non-tender firm abdomi-

Manuscript accepted for publication October 26, 2015

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doi: http://dx.doi.org/10.14740/jem318w

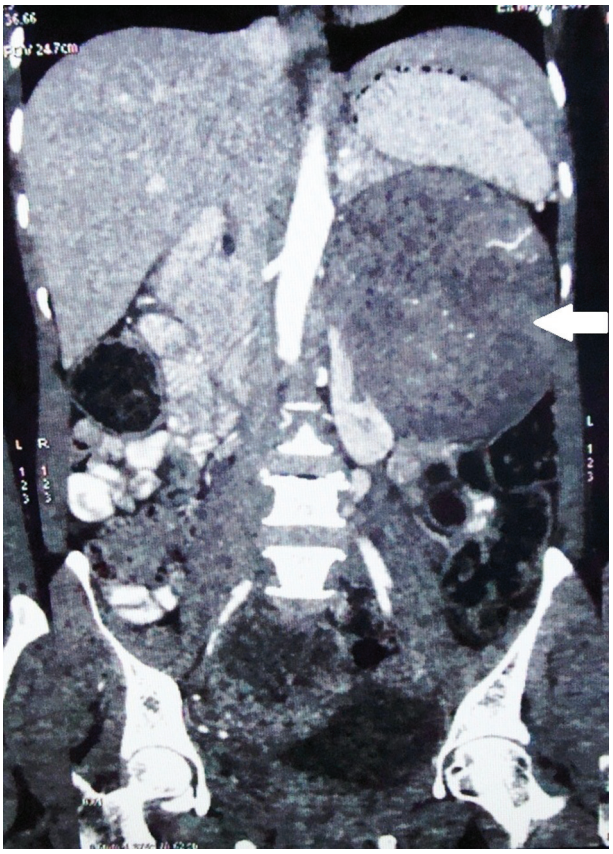

Figure 1. Contrast-enhanced computed tomography (CT) of abdomen showing a large left-sided adrenal mass measuring 12.2 × 10.3 × 8.0 cm.

nal mass with smooth and regular margin and moving with respiration. Screening ultrasonography revealed a retroperitoneal suprarenal mass suggestive of adrenal origin. Clinical stigmata of Cushing's syndrome, Addison's disease or pheochromocytoma were absent. Biochemical investigations were significant for normal electrolytes, normal morning 9 am cortisol (17 µg/dL), post-overnight dexamethasone suppressed morning cortisol (1.6 µg/dL) and normal urinary metanephrines (12.2 µg in 24 h). Plasma renin activity and serum aldosterone were not evaluated as the patient did not have hypertension. Serologic test for human immunodeficiency virus and Epstein-Barr virus (EBV) was negative. CT of abdomen revealed a large (12.2 × 10.3 × 8.0 cm) heterogeneously enhancing adrenal mass with regular margins abutting the upper pole of the left kidney, with areas of necrosis (Fig. 1). There was no internal calcification. The pre-contrast, immediate post-contrast and delayed post-contrast Hounsfield unit (HU) values of the adrenal mass were 65, 96 and 84, respectively. Hence the absolute and relative contrast washout values were 38.7% (> 60% suggestive of benign lesion) and 12.5% (> 40% suggestive of benign lesion) respectively, suggestive of malignant lesion [22]. Hence left adrenalectomy was done. The resected specimen weighed 91 g and measured 12 × 10 × 8.0 cm (Fig. 2A, B). Macroscopically, the tissue had a white appearance and there was no bleeding, ulceration or necrosis (Fig. 2A, B). Histopathology revealed a well-circumscribed and encapsulated benign spindle cell arranged in fascicles and whorls. There were few areas of hyaline degeneration. No features of mitosis or pleomorphism were present (Fig. 3A, B). Recovery was uneventful and in the last evaluation 6 months after the surgery, the patient was doing well, clinically asymptomatic, and had regained 8 kg body weight.

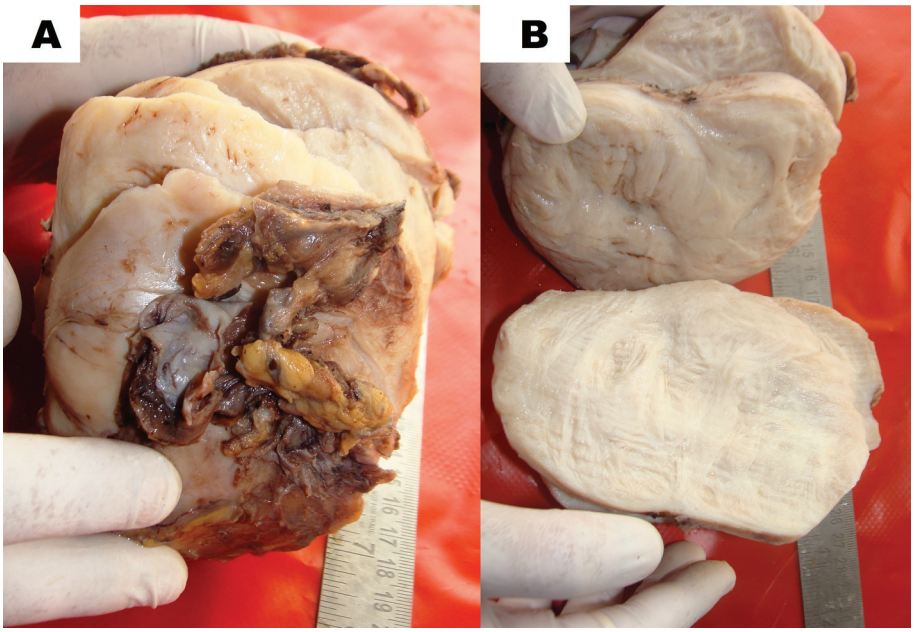

Figure 2. (A) Post-surgical resected specimen showing well-encapsulated adrenal mass. (B) Resected specimen's cut surface showing typical whorling appearance classical of leiomyoma.

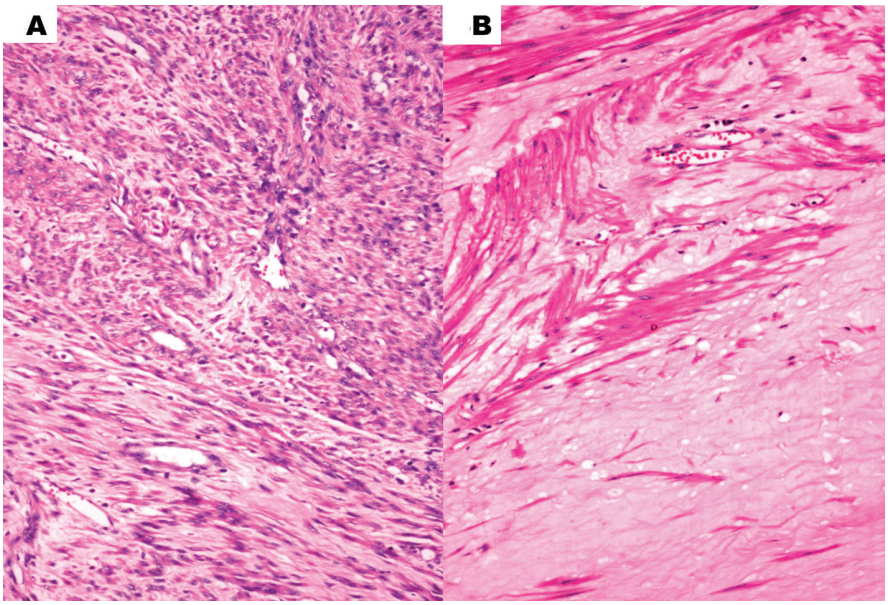

Figure 3. (A) Histopathology of the resected adrenal tumor showing bundles of smooth muscle cells diagnostic of leiomyoma (hematoxylin & eosin stain, × 200). (B) Higher magnification showing hyaline degeneration within the leiomyoma (hematoxylin & eosin stain, × 400).

Table 1. Profile of Patients With Adrenal Leiomyoma Reported in Literature as Compared to the Present Report

S. No.	Reported cases	Age (years)	Sex	Tumor characteristics		HIV status	EBV status
				Functional[a]	Size (mm)[b]		
1	Lin et al [1]	31	Female	No	110 × 90 × 70	Positive	Negative
2	Nishida et al [2]	48	Female	No	55 × 45 × 50	Negative	Negative
3	Alteer et al [3]	40	Male	No	49 × 55 × 60	Positive	Positive
4	Chang et al [4]	53	Female	No	55 × 45 × 35	Positive	Negative
5	Radin et al [5]	28	Female	NA	30	Positive	Negative
6	Jimenez-Heffernan et al [6]	2	Male	NA	70 × 50 × 50	Positive	Positive
7	Mouchet et al [7]	10	Female	NA	50 × 40 × 30	Negative	Negative
8	Gibbs et al [8]	49	Female	No	30 × 35 × 20	Negative	Negative
9	Jacobs et al [9]	65	Female	No	50 × 31 × 42	Negative	Negative
10	Parola et al [10]	35	Female	NA	35	Positive	Negative
11	Goldman et al [11]	72	Male	No	90 × 70 × 60	Negative	Negative
12	Dahan et al [12]	32	Male	No	30	Positive	Negative
13	Rosenfeld et al [13]	11	Female	NA	L: 50, R: 30	Positive	Negative
14	Demirel et al [14]	15	Male	No	L: 40 × 50 × 35 R: 80 × 50 × 30	Negative	Negative
15	Chao et al [15]	40	Female	No	NA	Negative	Negative
16	Jurczak et al. [16]	56	Male	No	72	Negative	Negative
17	Al-Masri et al [17]	38	Female	NA	NA	Negative	Negative
18	Parelkar et al [18]	11	Female	No*	L: 43 × 35 × 30 R: 90 × 70 × 40	Negative	Positive
19	Present report	42	Female	No	120 × 100 × 80	Negative	Negative

[a]Functional refers to the functional status of the tumor, viz. its ability to secrete one or more hormones either from the adrenal cortex or medulla. [b]For tumors with single value, it represents the largest diameter. *The 24-h urinary vanillylmandelic acid was normal. Plasma normetanephrine was increased, which is commonly known to be false-positive. NA: not available; HIV: human immunodeficiency virus; EBV: Epstein-Barr virus; L: left; R: right.

Discussion

Leiomyomas have been most commonly reported from the uterus followed by the gastrointestinal tract [1-3]. Leiomyomas are extremely rare tumors of the adrenal gland [1-5]. They are classically asymptomatic, and usually detected incidentally when abdominal imaging is done for some other cause (incidentaloma), and diagnosis is usually on histopathologic examination of the resected specimen. The cases reported in literature have been summarized in Table 1 [1-18]. Leiomyomas have a varied age of presentation ranging from 2 to 72 years (median: 38 years) with a female preponderance (12 out of 18; 66.67%) (Table 1). Adrenal leiomyomas reported in literature were typically unilateral, solitary and have a varied size ranging from 30 to 110 mm in diameter (median largest diameter: 50 mm) (Table 1). Bilateral adrenal leiomyoma are even rarer, predominantly reported in children [14, 18]. We would like to highlight that the leiomyoma in our patient is the largest adrenal leiomyoma ever reported in indexed medical literature. Interestingly immunodeficiency has been linked to adrenal leiomyoma both in the pediatric and adult age groups. HIV infection or acquired immune deficiency syndrome (AIDS) has been observed in eight out of 18 (44.44%) cases reported [1-18]. There are few reports of adrenal leiomyoma occurring in patients with latent EBV infection (three out of 18; 16.67%) [3, 6, 18]. However, direct link between immunodeficiency and adrenal leiomyoma has not been established. It has been postulated that HIV infection may promote smooth muscle tumors through a direct or indirect oncogenic stimulatory effect [10]. HIV is well known to be associated with lymphomas and Kaposi sarcoma [10]. All the adrenal leiomyomas reported till date (including our patient) were biochemically non-functional, viz. did not have any evidence of one or more hormone production either from adrenal cortex or medulla.

It is important to highlight that in contrast to previous reports, our patient did not have immunodeficiency, and was clinically symptomatic at the time of presentation. The radiologic features on CT in our patient mimicking malignancy have also been observed previously [1, 12, 18]. This report intends to highlight that adrenal leiomyoma is a rare benign non-functional adrenal neoplasm with good clinical outcomes following surgical excision. Adrenal leiomyoma can present as huge abdominal masses mimicking malignancy, and even radiologic investigations can be misleading in terms of differentiating this benign lesion from malignant adrenal lesions like adrenal carcinoma or pheochromocytoma. Adrenal leiomyoma should always be considered in the differential diagnosis of huge unilateral non-functional incidentally detected adrenal lesions.

Conflict of Interests

None.

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