

Case Report

Leiomyoma of the Adrenal Gland Presenting as An Incidentaloma

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Abstract

This was an unusual case of a primary leiomyoma tumor of the adrenal gland in a 26-year-old woman incidentally found during workup for mild, dull abdominal pain.

Keywords: adrenal gland, adrenal incidentaloma, leiomyoma, pathology

Introduction

Adrenal incidentalomas are clinically unapparent adrenal masses discovered inadvertently in the course of diagnostic testing or treatment for other clinical conditions that are not related to adrenal disease.¹ Their incidence has risen recently due to improved imaging studies and their increased use.^{2,3} The differential diagnosis of incidentaloma consists of a long list of rare, miscellaneous tumors, including leiomyoma.⁴

In this paper, we report a case of adrenal leiomyoma suspicious for either hepatic or hydatid cyst pre-operatively. A literature review of previously reported adrenal gland leiomyomas is also presented.

Case Report

A 26-year-old woman presented with a history of dull, mild abdominal pain in the right upper abdominal quadrant since six months previous, with aggravation during the last month. The pain worsened with eating and lasted for about 2 hours. She also had a history of weight loss and anorexia but not early satiety or nausea and vomiting. She was a known case of hypothyroidism who was treated with levothyroxin therapy (100 µg orally per day). Liver function tests, hematologic workup and 24 hour urine for Vanillyl mandelic acid (VMA) and catecholamines measurements were within normal limits. HIV antibody assay via the ELISA technique was negative.

Physical exam revealed a normotensive non-icteric woman with no palpable abdominal mass.

Abdominal sonography revealed a hetero-echo 9 cm diameter mass with a calcified peripheral wall on the right adrenal gland. Computed tomography (CT) scan was also performed which showed a heterogeneous right adrenal mass (9×7 cm) with multiple hypo-attenuated foci (Figure 1). Radiological differential diagnosis included adrenal cortical adenoma, carcinoma, and pheochromocytoma.

Chest X-ray was performed and according to normal findings of this evaluation, no more radiological studies (i.e., chest CT scan) were ordered.

Surgery was successful with no adhesion of the mass to neighboring tissues. The mass, in its entirety, plus the partially adhered right adrenal tissue were removed. The patient had an uncomplicated recovery and was discharged from the hospital three days after admission. There was no long-term follow up of the patient.

Gross pathologic evaluations indicated a cream-orange smooth, round, calcified mass that measured 9×7×5 cm and weighed 285 g. The mass was hard and could not be dissected by a knife, thus it initially was dissected by saw, then embedded in nitric acid (10%) for 24 hours. Microscopic evaluations showed interlacing bundles of bland smooth muscle cells without mitotic activity (Figure 2B). Foci of calcification and new bone formation were evident in the stroma. Residual adrenal tissue was also noted at the periphery of the smooth muscle cells (Figure 2A). The diagnosis of leiomyoma was supported by immunohistochemical (IHC) studies for smooth muscle actin (SMA), which was diffusely intracytoplasmically positive in the benign-appearing smooth muscle tumor cells (Figures 2C and 2D).

Additional IHC studies included CD10, CD34, CD117, estrogen receptor (ER), progesterone receptor (PR), and anaplastic lymphoma kinase 1 (ALK-1) which were all negative.

Discussion

Primary leiomyoma of the adrenal gland is a rare tumor associated with the adrenal vein or its tributaries.⁵ Only 14 cases have been previously reported in the literature (Table 1) with an age range from 2 to 72 years (average: 36 years).⁶⁻¹⁸ Adrenal leiomyoma predominates in females (10 cases out of 15; 66.6%). Only two cases had bilateral masses and seven, as with our case, were right sided. Tumor sizes ranged from 0.7 to 11 cm (average: 6.2 cm). Adrenal weight was between 17.8 and 315 g.⁴ The current case had a maximum diameter of 9 cm and 285 g weight.

Adrenal incidentalomas are defined as adrenal tumors discovered by imaging studies performed for other conditions.¹ Leiomyomas are regarded as one diagnosis in the long list of differential diagnoses for incidentalomas. They occur most frequently in the uterus and gastrointestinal tract but can be identified in different places, including the adrenal glands.¹⁶

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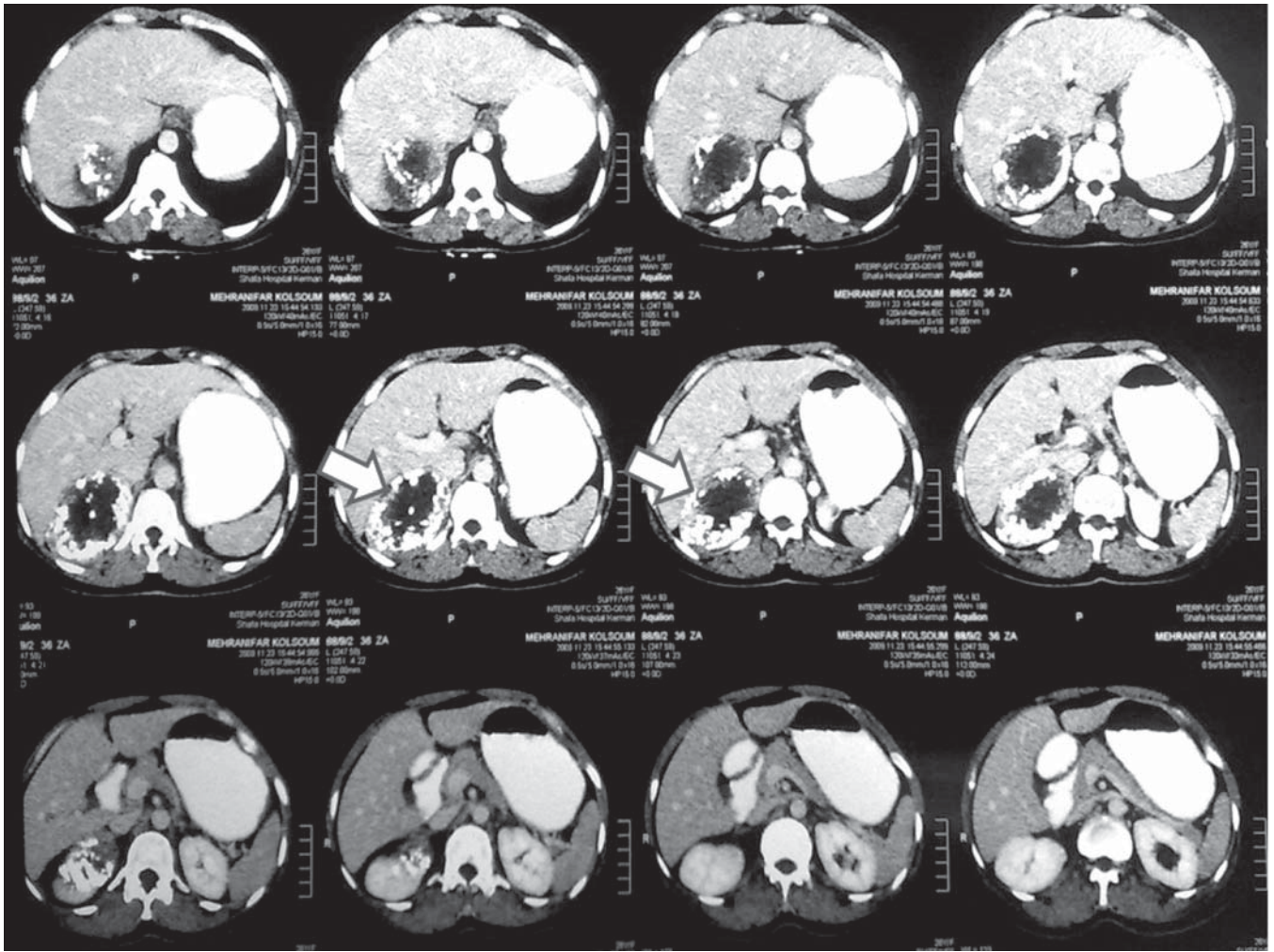


Figure 1. Abdominal computed tomography (CT) scan showed a heterogeneous right adrenal mass with multiple hypo-attenuated foci (arrow-heads).

Table 1. Reported cases of adrenal leiomyoma.⁴

Report	Size (cm)	Weight (g)	Age (yr)	Sex	Side	Functional	Clinical associations	Reference
1	5.5×4.5×3.5	Un	53	F	Left	No	None	6
2	3×3.5×3	17.8	49	F	Left	No	AIDS(+), EBV(-)	10
3	5×3.1×4.2	Un	65	F	Left	No	None	12
4	3.5	Un	35	F	Left	Un	Un, AIDS(+)	17
5	7×5×5	Un	2	M	Right	Un	AIDS(+), EBV(+)	13
6	5.5×5×4.5	84	48	F	Left	No	Hypertension	16
7	9×7×6	Un	72	M	Left	No	None	11
8	3×3×3	Un	32	M	Left	No	AIDS	8
9	5×4×3	Un	10	F	Left	Un	Ataxia-telangiectasia	15
10	L: 5; R: 3	Un	11	F	Bilateral	Un	AIDS	18
11	L: 4×5×3.5; R: 8×5×3	L: 24.5; R: 80.5	15	M	Bilateral	No	Multiple Leiomyomas involving spleen, epicardium, bilateral testicular microlithiasis, and empty sella tursica	9
12	Un	Un	40	F	Right	No	None	7
13	7.2	Un	56	M	Right	No	None	14
14	11×9×7	315	31	F	Right	No	None	4

Un=unknown; R=right; L=left; AIDS=Acquired immune deficiency syndrome; EBV=Epstein-Barr virus.

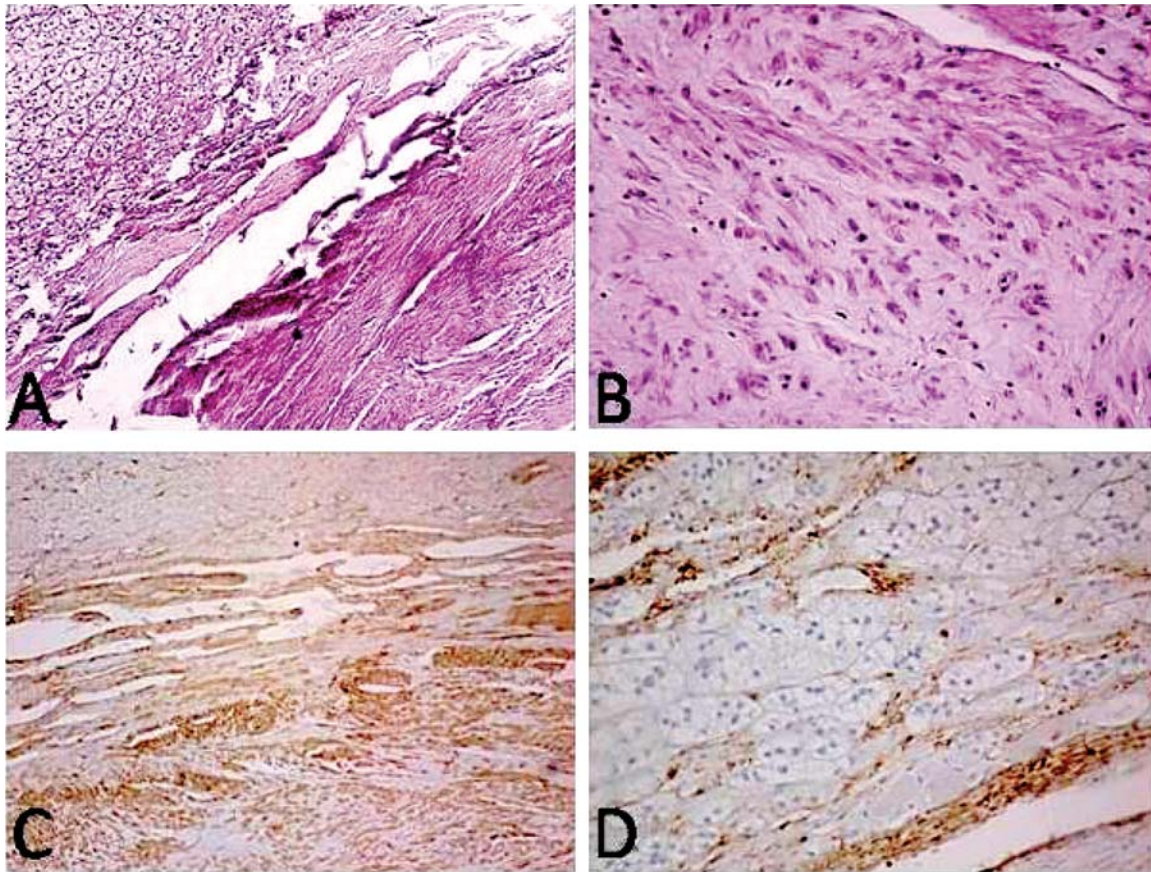


Figure 2. **A)** Microscopic view of adrenal leiomyoma shows tumor cells at the lower right side of the slide and normal adrenal tissue at the upper left side (H&E, 100 \times). **B)** Section shows interlacing bundles of smooth muscle cells (H&E, 400 \times). **C)** Immunohistochemical (IHC) staining for smooth muscle actin (SMA) revealed a strong intracytoplasmically positive reaction (100 \times). **D)** Note the negative reaction to SMA staining in the normal adrenal cortical tissue (400 \times).

The adrenal leiomyomas are usually small tumors, however, as in this case, they can manifest a pressure effect on the neighboring organs. In addition to hematoxylin and eosin (H&E) staining, among IHC assays the tumor was positive for SMA, thus confirming the smooth muscle origin of this tumor. However, other benign tumors of smooth muscle origin, particularly inflammatory myofibroblastic tumors should be ruled out. The latter also has an inflammatory component rich in lymphoplasmic cells, which was absent in this case. In addition, other IHC markers such as CD10, CD34, CD117, ALK-1, ER, and PR were assessed, of which all were negative in this case. Inflammatory myofibroblastic tumors are positive in 50% of cases^{17,18} and are sometimes positive for CD117.^{19,20} Additional IHC stains have also been used by some authors [i.e., S100, desmin, epithelial membrane antigen (EMA), CD34, Bcl-2, and CD117 (c-kit)].⁴ This case is 10 years younger than the average age of the reported cases.

As seen in Table 1, many reported cases of adrenal leiomyomas co-exist in patients diagnosed with human immunodeficiency virus/acquired immunodeficiency syndrome (HIV/AIDS) and/or latent Epstein-Barr virus (EBV) infection.¹³ Among the reported cases of adrenal leiomyoma, five are from AIDS patients.^{8,10,13,21,22} The association of leiomyoma and HIV/AIDS is not restricted to the adrenal gland, as these tumors have been found in various sites in children and adults.^{23,24} According to ELISA, our case was HIV-

negative. EBV infection has been associated with smooth muscle tumors in immunocompromised patients but not in immunocompetent patients.²³ The pathologic reason for this association is not clear. The EBV status of our case was not clear.

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A view of Latyan dam which is located around 25 km from Tehran. (Photo by M.H. Azizi MD)