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Case Report

Primary adrenal leiomyosarcoma: a case report and review of literature

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Abstract: Primary adrenal leiomyosarcoma (PAL) is an extremely rare mesenchymal tumors and originates from the smooth muscle wall of the central adrenal vein and its branches. Herein we report a case of a 49-year-old female suffering from PAL. Computed tomography revealed a well-circumscribed heterogeneously mass measuring 6×5×5 cm located in the left suprarenal areal, and a left laparoscopic adrenalectomy was underwent. Microscopic examination showed a hypercellular tumor with intersecting fascicled of spindled cells. Immunohistochemical staining showed that the cells were positive for desmin, smooth muscle actin (SMA), vimentin and negative for CD34, CD117, S100, Bcl-2 and Dog1. No oncological treatment underwent after surgery, and the patient had no recurrence or metastasis at 6 months postoperatively.

Keywords: Adrenal gland, adrenal gland neoplasms, leiomyosarcoma

Introduction

Leiomyosarcoma is a soft tissue neoplasm of smooth muscle origin. Although it may occur primarily in myometrium, retroperitoneum or dermis of the extremities, primary leiomyosarcoma of the adrenal gland is very rare. To the best of our knowledge, it has been reported only in 30 patients in the English language literature. In the present case, we report another case of primary adrenal leiomyosarcoma and review the clinical and pathological characteristics.

Case report

A 49-year-old female admitted presented with a 10-year history of left abdominal and back pain. The patient denied any other constitutional symptoms, as well as recent weight loss. She had a history of hypertension (160-170/90-100 mmHg), and her HIV antibody was negative. Physical examination showed no abnormal findings, except for mild abdominal discomfort at palpation. A computed tomography was performed, which revealed a well-circumscribed heterogeneously mass measuring 6×5×5 cm located in the left suprarenal areal (**Figure 1**). Neither venous thrombosis nor metastatic lesion was noted. 24 hour urine collections for cortisol and catecholamines were normal, as were serum aldosterone and ACTH levels. Based on a clinical diagnosis of nonfunctional adrenal tumor, a left laparoscopic adrenalectomy was conducted.

Gross pathological examination showed a roundish, grayish-white mass measuring 8×6×5 cm. The tumor tightly abutted the stretched identifiable adrenal gland. There were areas of hemorrhage and necrosis. The microscopic examination revealed a hypercellular tumor with intersecting fascicled of spindled cells (**Figure 2A**). Nuclear enlargement and occasional giant cells were noted. Resection margins were free of disease.

On immunohistochemical studies, the tumor cells stained positive for desmin (**Figure 2B**), smooth muscle actin (SMA) (**Figure 2C**), vimentin (**Figure 2D**) and negative for CD34, CD117, S100, Bcl-2 and Dog1. The proliferation rate ki67 was high (>60%). Based on the histopathological and immunohistochemical findings, the

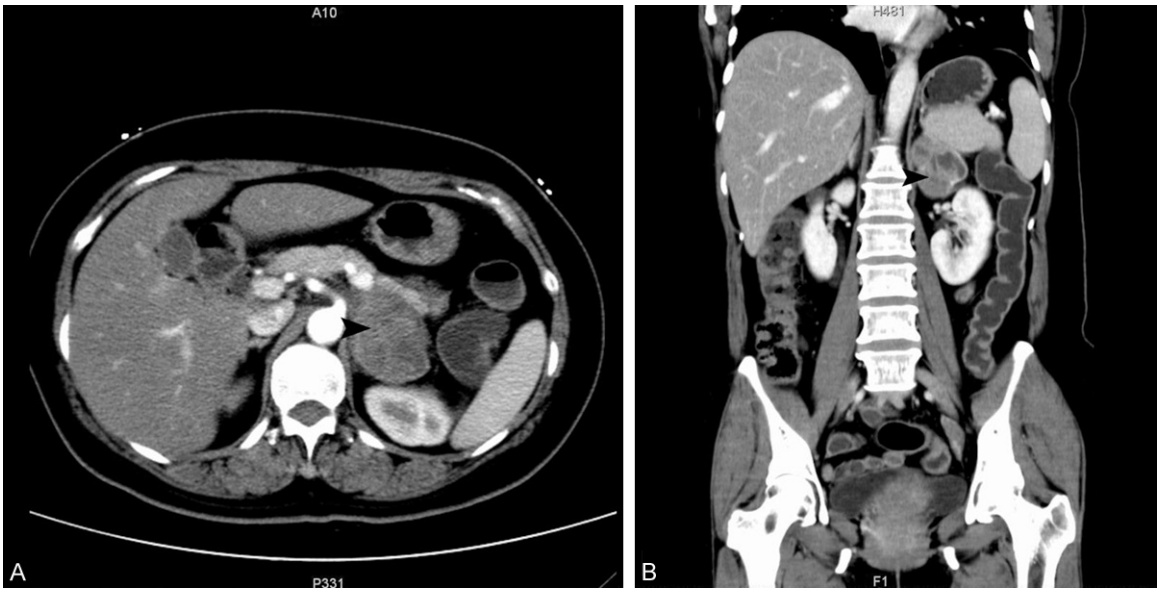

Figure 1. Computed tomography showed a well-circumscribed heterogeneously mass in the left suprarenal areal (arrowhead). (A) Axial sections and (B) coronal sections.

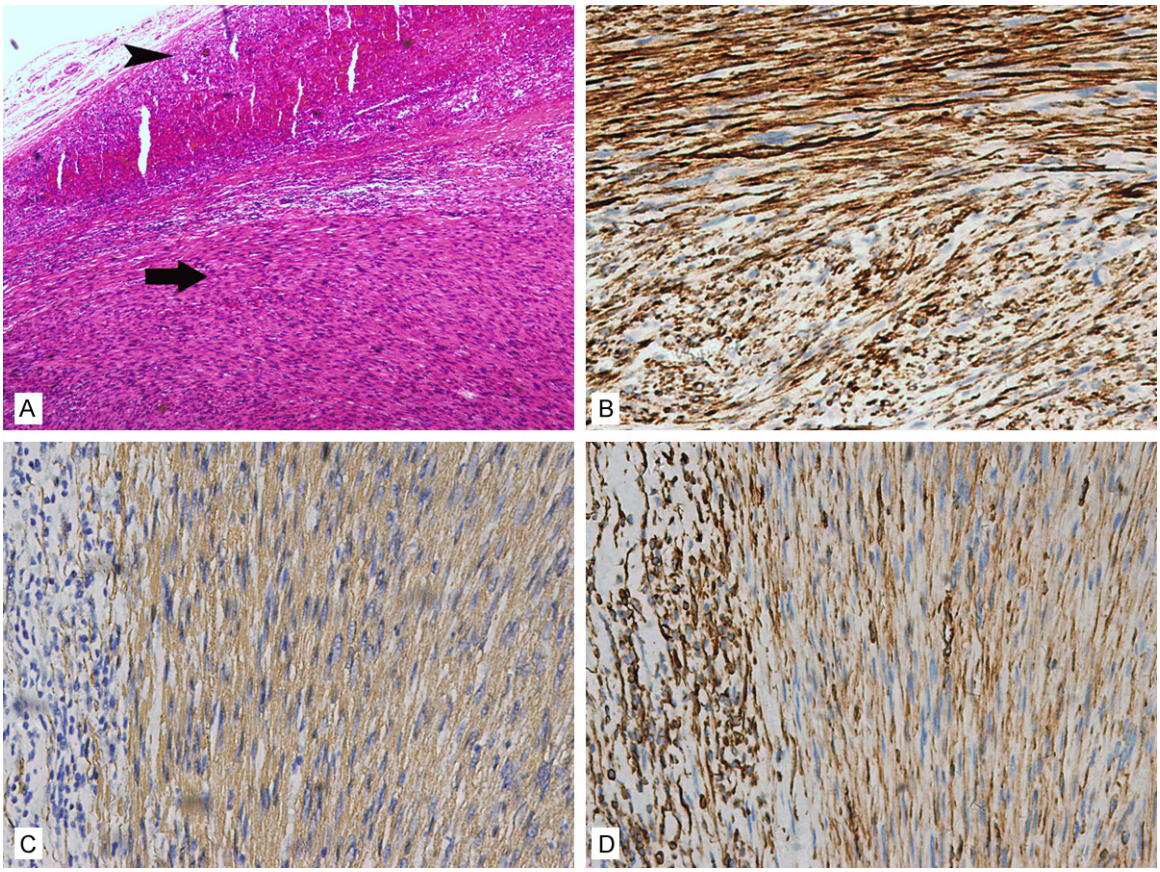

Figure 2. A. Hematoxylin and eosin stain shows the tumor (arrow) compressed the adrenal gland (arrowhead) (magnification ×40). B. Immunohistochemical staining for SMA is positive (magnification ×200). C. Immunohistochemical staining for desmin is positive (magnification ×200). D. Immunohistochemical staining for vimentin is positive (magnification ×200).

Table 1. Summary of previously reported cases of primary adrenal leiomyosarcoma

References	Age/y	Sex	Side	Size/cm	Extension	Treatment	Follow-up/months	Pathological Features
Choi. [1]	50	F	L	16	None	Adx + partial Nx	12 (alive without recurrence or metastasis)	ND
Lack. [2]	49	M	R	11	None	Adx + Nx + RT + CT	9 (alive with metastasis)	vimentin/actin/SMA (+)
Zetler. [3]	30	M	L	11	ND	Adx	20 (alive without recurrence or metastasis)	SMA (+)
Boman. [4]	48	M	R	2	ND	None	ND	SMA/HHF35/vimentin/desmin (+)
	29	M	L	0.8	ND	None	ND	SMA/HHF35 (+)
Etten. [5]	73	F	R	27	IVC	exploratory laparotomy	3 weeks (dead)	SMA (+)
Matsui. [6]	61	F	R	ND	IVC+right atrium	Adx + Nx + thrombectomy	1 (dead with metastasis)	SMA (+)
Lujan. [7]	63	M	R	25	Pulmonary metastasis and Invasion to AO	CT + Adx + Nx + hepatic lobectomy + cholecystectomy	died shortly after surgery	Pleomorphic.
Thamboo. [8]	68	F	R	13	None	Adx + Nx	12 (alive without recurrence or metastasis)	SMA/vimentin/actin/desmin (+)
Linos. [9]	14	F	Bil	3.5 (R) 4 (L)	None	Bil Adx	ND	SMA/viminten/actin/ HHF (+)
Kato. [10]	59	M	L	10	IVC	Adx + Nx + thrombectomy	6 (dead with metastasis)	Pleomorphic. SMA/desmin/vimentin (+)
Wong. [11]	57	M	L	ND	IVC and both iliac veins	Adx + Nx + thrombectomy	more than 6 months (dead with recurrence)	ND
Candanedo- Gonzalez. [12]	59	F	L	16	Invasion to AO	Adx + CT + RT	36 (alive with metastasis)	Pleomorphic. Actin/desmin/vimentin (+)
Lee. [13]	49	M	L	3	None	Adx	10 (alive without recurrence or metastasis)	desmin (+)
Mohanty. [14]	47	F	L	10	None	Adx + Nx + RT	9 (alive with metastasis)	Pleomorphic. desmin/calpinin/actin (+)
Wang. [15]	64	F	R	14	IVC + right atrium	Adx + thrombectomy	10 (alive without recurrence or metastasis)	SMA/desmin (+)
Goto. [16]	73	F	R	8	Invasion to AO	Adx + Nx	10 (alive without recurrence or metastasis)	SMA/NSE (+)
Mencoboni. [17]	75	F	R	8	None	Adx	12 (alive without recurrence or metastasis)	SMA/desmin/actin (+)
Van Laarhoven. [18]	78	M	L	ND	multiple metastasis	RT	11days (dead with metastasis)	SMA/actin/vimentin (+)
Hamada. [19]	62	F	Bil	8 (R) 4 (L)	None	Bil Adx + CT + RFA + RT	16 (dead with metastasis)	SMA (+)
Karaosmano-glu. [20]	63	M	R	ND	IVC	CT	3 (dead)	actin/vimentin/desmin/keratin (+)
Shao. [21]	66	M	L	10	Renal vein	Adx	18 (alive without recurrence or metastasis)	SMA/desmin (+)
Kanthan. [22]	28	F	L	16	None	Adx + Nx + partial diaphragmatic	ND	Pleomorphic. SMA/vimentin (+)
Deshmukh. [23]	60	F	L	5	None	Adx	21 (alive without recurrence or metastasis)	SMA/vimentin/desmin (+)
Gulpinar. [24]	48	M	r	11	None	Adx	8 (alive without recurrence or metastasis)	SMA/vimentin (+)
Ozturk. [25]	70	F	R	8	IVC	Adx + cavatomy + CT	6 (alive with metastasis)	SMA/desmin (+)
Lee. [26]	28	M	R	15	None	Adx	18 (alive without recurrence or metastasis)	SMA/desmin (+)
Bhalla. [27]	45	M	R	11	multiple meta	CT	9 (alive with metastasis)	desmin/actin (+)
Wei. [28]	57	F	L	8	None	Adx	29 (alive without recurrence or metastasis)	SMA/vimentin/actin/desmin (+)

Bil, bilateral; ND, not determined; IVC, inferior vena cava; AO, adjacent organ; Adx, adrenalectomy; Nx, nephrectomy; RT, radiation therapy; CT, chemotherapy; RFA, radiofrequency ablation; SMA, smooth muscle actin; NSE, neuronspecific enolase.

diagnosis of an adrenal leiomyosarcoma was made.

The postoperative period was uneventful, and no oncological treatment was undergone after surgery. Recurrence or metastasis of the primary tumor has not been detected at 6 months postoperatively.

Discussion

Primary adrenal leiomyosarcoma (PAL) is extremely rare, and was first described by Choi and Liu in 1981 [1]. It is believed to originate from the smooth muscle wall of the central adrenal vein and its branches [2]. To the best of our knowledge, only 30 cases of PALs have been reported in the English-language literature (summarized in **Table 1**).

In almost all clinically reported cases to date, they are elderly patients with large tumors. Most patients present with abdominal or flank pain, and some with lower extremity edema, spider angiomata when the tumor invasions to the inferior vena cava. PALs occur in females and males to approximately the same extent, and equally locate in the right and left adrenals. There are only two cases of bilateral PALs in all these reported cases [9, 19]. Most of the reported cases are of conventional type and only 5 cases are of pleomorphic variety [7, 10, 12, 14, 22].

It is interesting to note that four of the patients were immune-deficient due to HIV or Epstein-Barr virus infection [3, 4, 9]. It seems that PAL is likely to occur in an immunosuppressive situation, but nothing certain is known about the pathogenetic involvement of these viruses.

As PALs do not produce any adrenal hormonal derangement and grow rapidly, there is no applicable tumor marker or imaging characteristics available for making a preoperative diagnosis and all the cases were diagnosed after surgery or at necropsy. Encouragingly, Goto et al. [16] reported a case of neuronspecific enolase (NSE)-producing PAL. The level of serum NSE was markedly high preoperatively and NSE protein was massively expressed in the resected tumor. After surgery, serum NSE level became normal. It is suggested that serum NSE level could be a useful hallmark for the early detection for PAL. However, Kato et al. [10] found immunostaining for neuron-specific enolase (NSE) was negative in their case. So we need a further research to seek a suitable tumor marker.

Histopathological and immunohistochemical evaluation is indispensable not only for determining tumor type but also for differential diagnosis. Conventional leiomyosarcomas show strong immunoreactivity for smooth muscle markers such as smooth muscle actin and/or muscle specific actin in 90 to 95% of cases, and desmin in 70-90% of cases [14]. However, there is a marked variability in the expression of these markers in pleomorphic leiomyosarcomas. Oda et al. [29] reported that 37.5% of the pleomorphic leiomyosarcomas of various sites are desmin positive, 46.4% are muscle-specific actin positive, and 50% are smooth muscle actin positive. Malignant fibrous histiocytoma, malignant melanoma, malignant hemangioperistoma, angiosarcoma, liposarcoma, carcinosarcoma, rhabdomyosarcoma, adrenal invasion by a retroperitoneal leiomyosarcoma and metastatic tumors should be considered in the differential diagnosis of adrenal leiomyosarcomas.

Radical surgery is the mainstay of therapy, but the prognosis for PAL patients is not predictable. It's believed that patients with invasive diseases that include venous thrombosis, adjacent organ invasion, and distant metastases, the prognosis is extremely poor [6]. We found that in all these 29 cases, 12 patients who had no recurrence or metastasis were almost without invasive diseases and none of them underwent any adjuvant therapy such as chemotherapy or radiotherapy. Adjuvant therapy combined with surgery is often used for PAL patients with poor prognosis. In a systematic overview study, Strander et al. [30] showed that postoperative adjuvant radiation therapy was recommended for the treatment of locally advanced malignancy in soft tissue sarcomas. Radiation therapy and/or chemotherapy may be helpful to shrink the tumor and destroy the remaining tumor cells.

In conclusion, we presented a rare case of primary adrenal leiomyosarcoma with characteristic clinicophathological features. The characteristic findings of imaging studies, the histological features and immunohistochemical staining for desmin, SMA and vimentin are helpful for

the diagnosis and differential diagnosis of primary adrenal leiomyosarcoma. For the time being, early and complete surgical resection is the mainstay management.

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Disclosure of conflict of interest

None.

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