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CASE REPORT

Pleomorphic Leiomyosarcoma of the Adrenal Gland in a Young Woman: A Case Report and Review of the Literature

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Abstract: Leiomyosarcoma usually arises in the uterus, abdominal and urologic viscera, and walls of large and small blood vessels. However, primary adrenal leiomyosarcoma is extremely rare with only 39 cases previously reported in English-language literature. We report a case of a 29-year-old previously healthy woman with an incidentally found right adrenal-occupying lesion. CT scan revealed a right adrenal mass measuring 3.3×3.4 cm in size. The tumor was successfully removed by laparoscopic adrenalectomy. Postoperative histopathologic examination showed spindle cells arranged in interlacing fascicles with pleomorphism and a high mitotic rate. An immunohistochemical examination showed positive staining for SMA, desmin, vimentin and H-caldesmon, and the diagnosis of a well-differentiated adrenal leiomyosarcoma was established. The patient received no other oncological treatment after surgery and currently has no evidence of residual disease or tumor recurrence according to imaging follow-up.

Keywords: adrenal gland, adrenal gland neoplasms, leiomyosarcoma, adrenalectomy

Introduction

Leiomyosarcoma is a malignant mesenchymal tumor composed of cells showing distinct features of the smooth muscle lineage.[1] The most common anatomic sites are the uterus, abdominal and urologic viscera, and walls of large and small blood vessels. Occasionally, these tumors arise in bone, in somatic soft tissues and extremely rarely in the adrenal gland.[2,3] In 1981, Choi SH et al reported the first case of primary adrenal leiomyosarcoma (PAL). There are only 39 cases reported in English-language literature so far. Definitive diagnosis of PAL is usually achieved only after pathologic examination of the surgical specimen. Due to the development and wide use of imaging techniques such as computed tomography (CT) and ultrasonography, unexpected and special incidentaloma lesions can be detected more easily. Herein, we describe a case of a young woman with PAL without any discomfort. In addition, we report on our analysis of pathological features and corresponding data extracted from the 39 reported cases. Our aim is to share our experiences with rare tumors and discuss the epidemiology, anatomical localization, and diagnosis and current treatment strategies.

Case Report

A 29-year-old previously healthy Chinese woman incidentally found a right suprarenal mass by abdominal ultrasound on a preemployment examination. The patient

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denied systemic symptoms, pain, fever, anorexia, and other notable medical history. Physical examination did not reveal any significant abdominal tenderness, lymphadenopathy or other findings. Routine laboratory examinations were normal, including complete blood count, renal function and electrolyte levels. All the hormonal data showed that 24-h urine cortisol and plasma aldosterone to renin ratio were within normal limits, as were catecholamines and ACTH levels. Cortisol post 1 mg dexamethasone was 0.5 μg/dl (normal < 1.8). HIV-1 and HIV-2 antibody tests were non-reactive. Abdominal ultrasound showed the presence of a diffusely hypoechoic, homogeneous mass in the region of the right suprarenal area measuring 4.2×2.5 cm. We proceeded with computed tomography (CT) of the abdomen, and it revealed a well-defined tumor originating from the right adrenal gland that was 3.3×3.4 cm in size; the tumor, exerted pressure on the hepatic vein without evidence of regional adenopathies or infiltration of surrounding tissues. Contrast-enhanced CT showed moderate enhancement with irregular non-enhanced areas within, suggesting malignancy (Figure 1). A positron emission tomography/computed tomography (PET/CT) scan with ^{18}F-FDG was performed, which revealed an exclusive and high uptake in the right adrenal tumor lesion.

A right adrenalectomy was scheduled and performed uneventfully. Intraoperatively, we found a solid mass that almost completely replaced the right adrenal gland adherent to the posterior wall of the inferior vena cava (IVC). We explored the IVC and adjacent organs, and no tumor invasion was detected. Tissue margins were negative. Postoperatively her vital signs remained stable and as there were no complications. No adjuvant therapy and other medication was given, and the patient was discharged 2 days after surgery. Currently, she is alive and doing well, without evidence of recurrence or distant metastasis at the 12-month follow-up.

Gross pathological examination showed a well-circumscribed and partially encapsulated solid tumor weighing 37.5 g and measuring 5.5×5×3.2 cm in maximum dimension. The cut surface was nodular and grayish white in color with few mucoidal, hemorrhagic and necrotic areas. The normal adrenal gland was displaced by the tumor and presented at the edge of the tumor (Figure 2). Microscopically, the adrenal gland was compressed, but not invaded by the spindle cell tumor, which was arranged in interlacing fascicles (Figure 3A). Tumor cells were elongated with eosinophilic fibrillary cytoplasm with marked pleomorphism of the nuclei, with up to 8–10 mitoses/10 high power fields (Figure 3B). No infiltrated lymph nodes were found. An immunohistochemical examination showed positive staining for SMA, desmin (Figure 3C), vimentin, and H-caldesmon (Figure 3D), and negative staining for S-100, CD 117, Dog-1, ER and PR. The Ki-67 proliferation index was approximately 40% in the hot spot. Based on these data, the final diagnosis was confirmed as a primary adrenal leiomyosarcoma.

Discussion

PAL is an extremely rare nonfunctional mesenchymal tumor, and to the best of our knowledge, there have only been 39 previously published cases of PAL in English-language literature. The clinicopathologic characteristics of these cases are summarized in Table 1. PAL is mostly found in middle-aged adults. The age at first diagnosis ranges from 14 to 79 years, with a mean of 54.2 years.

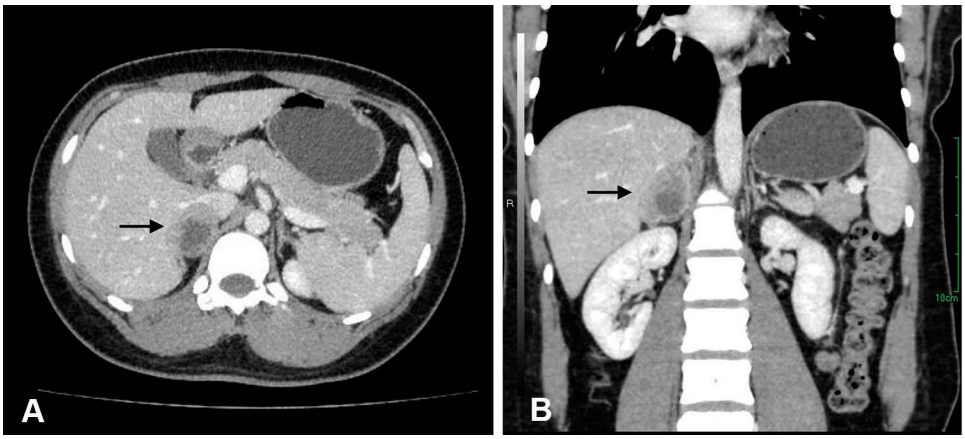

Figure 1 Preoperative abdominal contrast-enhanced CT scan showed a well-circumscribed heterogeneously mass in the right suprarenal areal (arrow). (**A**) Axial sections and (**B**) coronal sections.

Figure 2 Macroscopic features of the tumor showed a well-circumscribed and partially encapsulated solid tumor measuring 5.5×5×3.2 cm in maximum dimension. The normal adrenal gland was displaced by the tumor and presented at the edge of the tumor.

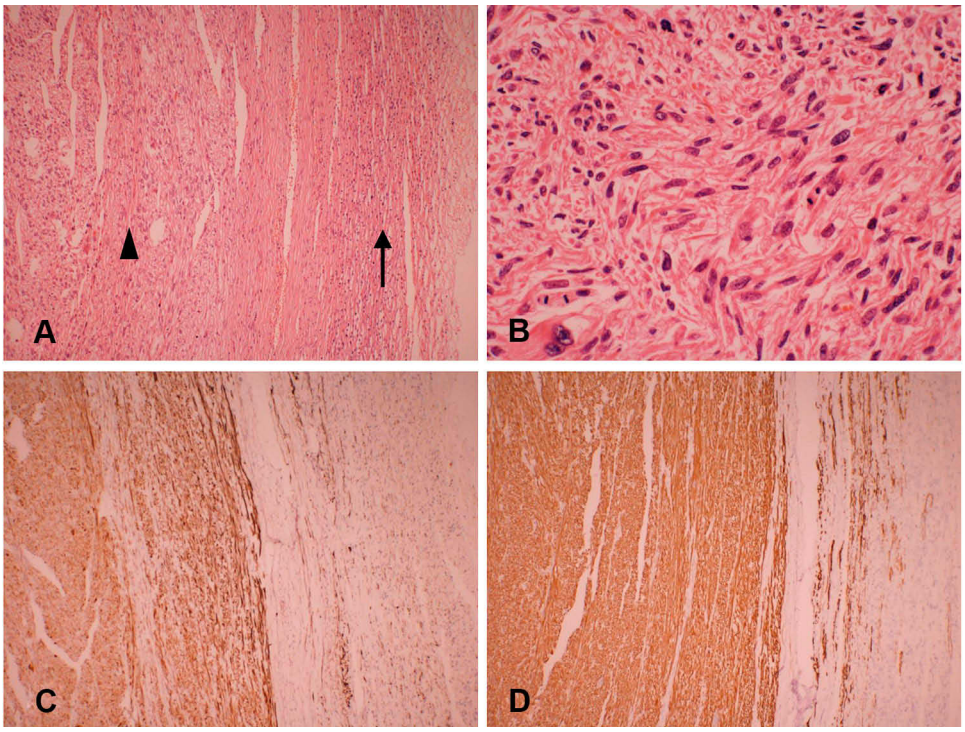

Figure 3 Microscopic details of the tumor. (**A**) The interlacing bundle and fascicles of the tumor (arrowhead) and compressed adrenal tissue (arrow). (H&E, × 100). (**B**) Leiomyosarcoma with nuclear pleomorphism and giant cell formation with mitotic activity in the range of 8–10 mitoses/10 high power fields (H&E, × 400). (**C**) Immunohistochemical staining for desmin is positive (× 100). (**D**) Immunohistochemical examinations showed strong immunoreactivity for H-caldesmon (× 100).

Table 1 Clinicopathologic Characteristics of Primary Adrenal Leiomyosarcoma

Author, Year	Age, y/Sex	Presentation	Side	Size (cm)	Treatment	Outcome (months)	Extension	Pathologic Characteristics of Known Cases of PAL in the Literature
Our case	29/F	Incidental finding	R	5.5	Adx	12m AWOD	None	Pleomorphic+ SMA/ desmin/vimentin/ H-caldesmon+
Sakellariou M et al, 2020[26]	62/M	Incidental finding	L	13	Adx; RT+CT	31m, alive with bone, lung and liver metastasis	None (preoperative); bone metastasis (postoperative after 3 months)	SMA/desmin+
Nerli RB et al, 2019[27]	27/M	Back pain	L	9	Adx	N/D	None	Desmin/H-caldesmon+
Doppalapudi SK et al, 2019[6]	70/M	Abdominal varices+EBLE	R	9	Adx + Nx+ thrombectomy+ cavatomy	14m, deceased	IVC (preoperative); lung metastasis (postoperative after 12 months)	SMA/desmin/vimentin/ H-caldesmon+
Mulani SR et al, 2018[28]	50/M	Abdominal pain + weight loss	L	8.1	CT+RT	N/D	Liver and lung metastasis	Pleomorphic+ desmin +
Onishi T et al, 2016[29]	34/M	Flank pain	R	5.2	Adx+ Nx+ Ldx+ partial hepatic lobectomy	10m AWOD	IVC+AO	Pleomorphic+ SMA+
Zhou Y et al, 2015[24]	49/F	Abdominal pain+ back pain	L	8	Adx	6m AWOD	None	SMA/desmin/vimentin+
Quildrian S et al, 2015[30]	44/F	Abdominal pain	R	12	Adx	36m, AWOD	None	Pleomorphic+ SMA/ desmin/vimentin/ H-caldesmon/HHF+
Nagaraj V et al, 2015[31]	61/M	Flank pain	L	17	Adx	N/D	None	Pleomorphic+ desmin/ vimentin+
Wei J et al, 2014[32]	57/F	Incidental finding	L	8	Adx; tumor resection +CT	29m, AWOD	None (preoperative); recurrence (postoperative after 17 months)	Pleomorphic+ SMA/ desmin/vimentin+
Oztürk H et al, 2014[33]	70/F	Flank pain	R	7.8	Adx+ cavatomy	6m, metastasis	IVC	Pleomorphic+ SMA/ desmin +
Lee S et al, 2014[34]	28/M	Flank pain +weight loss	R	15	Adx	18m, AWOD	None	Pleomorphic+ SMA/ desmin +
Gulpinar MT et al, 2014[35]	48/M	Frequent urination	R	11	Adx	8m, AWOD	None	SMA/vimentin+
Bhalla A et al, 2014[36]	45/M	Back and groin pain+weight loss	R	11	CT	9m, metastasis	Liver metastasis	SMA/desmin+
Alam MM et al, 2014[37]	35/F	Flank pain	L	8.5	Adx	N/D	None	ND

(*Continued*)

Table 1 (Continued).

Author, Year	Age, y/Sex	Presentation	Side	Size (cm)	Treatment	Outcome (months)	Extension	Pathologic Characteristics of Known Cases of PAL in the Literature
Deshmukh SD et al, 2013[17]	60/F	Flank pain	L	5.2	Adx	21m, AWOD	None	Pleomorphic+ SMA/ desmin/vimentin+
Shao IH et al, 2012[38]	66/M	Abdominal fullness + nausea	L	9.5	Adx + thrombectomy	18m, AWOD	Renal vein	SMA/desmin +
Karaosmanoglu AD et al, 2010[39]	63/M	Abdominal pain+EBLE	R	N/D	CT	3m, deceased	IVC	Desmin/vimentin/actin +
Kanthan R et al, 2012[40]	28/F	Abdominal pain	L	16.5	Adx+Nx+partial diaphragmatic resection	N/D	None	Pleomorphic+ SMA/ vimentin+; desmin-
Liu SV et al, 2012[7]	79/F	Subcostal pain	L	6.3	Adx	12m, AWOD	N/D	N/D
Van Laarhoven HWM et al, 2009[41]	78/M	Hemithorax pain	L	N/D	CT+RT	11days, deceased	Multiple metastases	SMA/vimentin/actin+
Hamada S et al, 2009[4]	62/F	Flank pain	B	R(8) L(4)	Bil Adx+CT+RFA+RT	16m, deceased	None (preoperative); multiple metastasis (postoperative)	SMA+
Mencoboni et al, 2008[22]	75/F	Abdominal pain+ loin pain.	R	8	Adx	12m, AWOD	N/D	Pleomorphic+ SMA/ desmin/actin+
Goto J et al, 2008[20]	73/F	Flank pain +progressive hypertension	R	8	Adx+Nx+cavotomy	10m, AWOD	AO	SMA/NSE +
Wang TS et al, 2007[8]	64/F	Non-productive cough+ EBLE	R	14.2	Adx+thrombectomy +cavotomy	10m, AWOD	IVC, right atrium	SMA/desmin+
Mohanty SK et al, 2007[18]	47/F	Flank pain	L	10	Adx+Nx+RT	9m, alive with lung and liver metastasis	None (preoperative); lung and liver metastasis (postoperative after 9 months)	Pleomorphic+SMA/ desmin/H-calponin/ actin+
Lee CW et al, 2006[42]	49/M	Flank pain	L	3	Adx	10m, AWOD	None	Desmin+
Wong C et al, 2005[9]	57/M	Groin pain +cold feet	L	N/D	Adx+particial Nx+ thrombectomy	6m, recurrence, deceased	IVC, iliac veins and right atrium	N/D
Candanedo-González FA, 2005[43]	59/F	Flank pain +weight loss	L	16	Adx	24m, AWOD	None (preoperative); recurrence and liver metastasis (postoperative after 12 months)	Pleomorphic+desmin/ vimentin/actin+

(*Continued*)

Table 1 (Continued).

Author, Year	Age, y/Sex	Presentation	Side	Size (cm)	Treatment	Outcome (months)	Extension	Pathologic Characteristics of Known Cases of PAL in the Literature
Linos D et al, 2004[5]	14/F	Incidental finding	B	R (3.5) L(4)	Bilateral Adx	14m, AWOD	None	SMA/vimentin/actin/ HHF35+
Kato T et al, 2004[44]	59/M	Flank pain +back pain.	L	10	Adx+Nx +thrombectomy+RT	6m, deceased	IVC+renal vein (preoperative); bone and liver (postoperative after 1 month)	Pleomorphic+ SMA/ desmin/vimentin+
Thamboo TP et al, 2003[21]	68/F	Loin pain +fever	R	12.5	Adx+Nx	12m, AWOD	None	Pleomorphic+ SMA/ desmin/vimentin/actin+
Lujan MG et al, 2003[23]	63/M	Enlarging RUQ mass	R	25	Preoperative CT+Adx +Nx+partial hepatic lobectomy+ cholecystectomy	Deceased shortly after surgery	Invaded AO+lung metastasis	Pleomorphic+ desmin/ vimentin/H-calponin +; SMA/actin -
Matsui Y et al, 2002[11]	61/F	Flank pain +fever	R	N/D	Adx+Nx +thrombectomy	1m, deceased	AO+IVC+right atrium	SMA+
Etten B et al 2001[10]	73/F	EBLE +abdominal pain	R	27	Palliative + supportive care	3 weeks, deceased	IVC+AO	SMA+
Boman F et al, 1997[45]	29/M	Autopsy	L	0.8	None	N/D	None	SMA/HHF35+
Zetler PJ et al, 1995[46]	30/M	Abdominal pain	L	11	Adx	20m, AWOD	None	Pleomorphic+ SMA/ actin+
Hayashi J et al, 1995[47]	55/F	Abdominal pain+fever	R	N/D	Adx+Nx	52m, AWOD	IVC+right atrium+hepatic vein	N/D
Lack EE et al, 1991[19]	49/M	Flank pain	R	11	Adx+Nx; CT+RT	9m, alive with bone metastases	None (preoperative); bone (postoperative after 3 months)	Pleomorphic+ SMA/ vimentin/actin+
Choi SH et al, 1981[48]	50/F	Flank pain	L	16	Adx+partial Nx	12m, AWOD	None	N/D

Abbreviations: Adx, adrenalectomy; AO, adjacent organ; AWOD, alive without disease; CT, chemotherapy; EBLE, edema in both lower extremities; IVC, inferior vena cava; Lymx, lymphadenectomy; NSE, neuron-specific enolase; Nx, nephrectomy; N/D, not disclosed; PAL, primary adrenal leiomyosarcoma; RFA, radiofrequency ablation; RT, radiation therapy; RUQ, right upper quadrant; SMA, smooth muscle actin.

Of these patients, 77.5% have been ≥ 40 years of age. PAL is evenly distributed between the sexes with cases involving 21 women and 19 men. In addition, there does not appear to be any laterality preference, as right-sided and left-sided tumors occur with similar frequency. There have only been two cases of bilateral lesions,[4,5] but it is not known whether the bilateral tumors were both primary or if one had metastasized to the other adrenal gland.

Symptoms are mainly secondary to the mass effect of the tumor and local invasion. The most common presenting symptom is flank pain or abdominal pain in approximately 64% of patients. If the tumor extends into the IVC or the large tumor compresses the IVC, patients may present with IVC involvement, and experience obstructive symptoms such as lower extremity edema, spider angiomata, abdominal varices, and even altered sensation in the lower limbs.[6–10]

In a rarely case, the tumor ruptured suddenly, and the patient presented with severe and acute abdominal pain.[11] Because tumors are located in the retroperitoneum, and are often nonfunctional, they often grow to large sizes prior to noticeable symptoms. The size of the PAL at presentation has ranged from 0.8 to 27 cm (mean, 10.2 cm), with 78.4% of tumors larger than 6 cm.

Adrenal masses detected incidentally during radiological investigations account for 4%.[12] Although there are no definite features that can be used to diagnose PAL, imaging examination is useful for identifying the tumor size, location and metastasis, and evaluating its extent in relation to its surrounding structures, as well as distinguishing between potentially benign and malignant lesions. In a series of 705 patients with adrenal incidentalomas, it was demonstrated that unenhanced CT attenuation >10 and size greater than 7 cm had a high sensitivity for malignancy.[13] There is a growing consensus, however, that the size should not be used to differentiate malignant from benign adrenal tumor. Modern imaging techniques such as magnetic resonance imaging (MRI) and ^{18}F-FDG-PET represent the state of the art in adrenal imaging, which have greater sensitivity and specificity in establishing malignant versus benign diagnosis. MRI provides a more precise estimation than CT in the detection of the tumor origin and location to local structures due to its high-quality soft tissue resolution. Generally speaking, malignant masses are denser than benign masses, and often appear hyperintense on T2-weighted images on account of their higher fluid content.[14] Multiplanar postcontrast imaging is also useful to evaluate the extent of vascular involvement and to distinguish tumor from bland thrombus.[15] Evaluation using FDG-PET has a high sensitivity for detecting malignancy; increased FDG accumulation can be seen in all cases of PAL that performed FDG-PET. Hamada et al reported that the metastasis was detected by PET before it was found by CT.[4] A core biopsy may be performed with radiological or ultrasound assistance prior to surgical resection. The current guidelines for the management of adrenal tumors recommend that the tumor should be confirmed to be hormonally inactive (in particular, a pheochromocytoma has been ruled out) prior to biopsy.[16] In addition, nonfunctional primary adrenal cortical carcinoma should also be considered as a differential diagnosis prior to an adrenal biopsy because tumor seeding can take place.

There is currently no tumor marker or imaging that can aid to distinguish PAL nodules from other adrenal nodules nor is there an identified characteristic genetic rearrangement or specific endocrinological change associated with this neoplasm. The diagnosis of PAL is still completely dependent on histological and immunohistological evaluations after surgery. Histologically, retroperitoneal smooth muscle tumors containing 5 or more mitoses per 50 high power fields are classified as malignant. The presence of tumor cell necrosis is also strongly suggestive of malignancy.[17] The typical histologic features of leiomyosarcomas include intersecting, sharply marginated fascicles of spindle cells with abundant eosinophilic cytoplasm and elongated and hyperchromatic nuclei. In many cases, coagulative necrosis and hemorrhage are common features. Varying degrees of nuclear hyperchromasia and pleomorphism usually manifest.[15] Pleomorphic leiomyosarcomas that resemble any undifferentiated soft tissue sarcomas show moderate-to-high anaplasia as in the index cases and need a panel of immunohistochemical stains to arrive at a definitive diagnosis.[18] Pleiomorphic subtypes are not uncommon with 18 known cases (45%). Some cases did not reveal pleiomorphism, but prominent atypical nuclei and bizarre giant cells were described. This implies that leiomyosarcoma should be considered in the cases of an adrenal mass with malignant spindle and pleomorphic cells; however, poor prognosis is not necessarily indicated with tumors with bizarre giant and pleomorphic cells.

Tumor cells are widely presumed to arise from the smooth muscle wall of the inferior vena cava, central adrenal vein, and its branches.[19] Direct infiltration of leiomyosarcoma cells was not observed microscopically in adjacent adrenal tissue in five cases. Some findings revealed that the tumor was surrounded by a thin rim of fibrous tissue and bordered the grossly identifiable adrenal gland. Therefore, we can further confirm that leiomyosarcoma most likely arises from adrenal blood vessels but not the adrenal gland.[8,20,21]

Immunohistochemistry is an extremely useful tool not only for determining tumor type, but also for differentiating leiomyosarcomas from other tumors (including melanoma, rhabdomyosarcoma, mesotheliomas and other sarcomas). Conventional leiomyosarcomas show strong reactivity for smooth muscle markers such as smooth muscle actin and/or muscle specific actin in 90–95% of cases and desmin in 70–90% of cases.[17] However, due to the lack of such detection in many cases of PAL, we cannot obtain reliable statistical data.

Surgical resection with wide negative margins constitutes the predominant mode of therapy,[16] and has the greatest impact on survival. Since tumors are typically large at diagnosis and likely to invade the IVC, it is important to carefully resect any possibly involved tissue.

It is worth noting that some patients develop pulmonary embolism after surgery, so we should strengthen this aspect of care for patients.[8,22]

The role of chemotherapy or radiation is not well defined. Although some reports suggested that radiation therapy could alleviate symptoms for patients with metastasis and ensure improved local control,[2] the sensitivity was generally poor. Kato T et al[21] performed palliative radiation therapy for the patient with cervical spine metastasis, but it showed almost no effect. Systemic chemotherapy may delay local recurrence if given as adjuvant/neoadjuvant treatment, but there are lack of randomized trial data indicating a survival benefit for patients.[2] In one case reported by Lujan MG et al,[23] the patient received preoperative chemotherapy, resulting in considerable reduction of the size of the lung mass; however, the adrenal mass continued to grow.

Given the rarity of the tumor, assessment of its prognosis remains difficult, but venous thrombosis, adjacent organ invasion and distant metastases were proposed to be predictors of an unfavorable outcome.[7,24] A prospective study suggested that 55% of patients with leiomyosarcomas of somatic soft tissues developed metastasis or died within 3 years.[25] We found that of the 40 patients, 7 died within six months. Because only limited clinical data are currently available on PAL, our data offer some important information on the clinical and therapeutic aspects of this rare disorder to increase clinician vigilance.

Conclusion

In conclusion, PAL is a rare mesenchymal neoplasm that most likely arises from the IVC, central adrenal vein and its branches. The tumors may grow to large sizes before incidental detection at imaging or palpation. Patients presenting with symptoms from these tumors might have large tumors that have invaded adjacent structures or extended into the IVC. Diagnosis in an advanced stage contributes to a poor prognosis. Surgical resection with clear margins remains the mainstay of treatment.

Ethical Approval

This study was approved by the ethics committee of the First Hospital of Jilin University (Changchun, People's Republic of China), and was permitted to be published. Written informed consent to have the case details and accompanying images published was obtained from the patient. All clinical investigations were conducted in accordance with the principles expressed in the Declaration of Helsinki.

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Dr. Yuanyuan Wang and Dr. Yongliang Teng are co-first authors for this study.

Disclosure

The authors declared no conflicts of interest.

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