Case Report

Leiomyosarcoma of the Abdominal Aorta: A Rare Cause of Renovascular Hypertension

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We describe the case of a 44-year-old woman who presented with renovascular hypertension caused by primary leiomyosarcoma of the abdominal aorta that had metastasized into the renal arteries. Despite an extensive radiological evaluation, the diagnosis was mistaken first for Takayasu's arteritis and then for retroperitoneal hematoma or neoplasm. The patient developed renal failure due to bilateral renal infarction, and died 3 months after her initial presentation with ischemic colitis. Postmortem examination confirmed the diagnosis. (*Hypertens Res* 2007; 30: 279–283)

***Key Words*: leiomyosarcoma, aorta, renovascular hypertension, retroperitoneal, CT scan**

Introduction

Leiomyosarcoma is a rare malignant tumor originating from the smooth muscle cells. Vascular leiomyosarcoma constitutes about 2% of all leiomyosarcomas and involves veins five times more than arteries (*1*, *2*). The diagnosis of vascular leiomyosarcoma is often missed because the clinical manifestations are non-specific and vary widely according to the location of the tumor, and because no reliable tumor marker is available. Unlike leiomyosarcoma of the inferior vena cava (*2–4*), aortic leiomyosarcoma is an extremely rare and very ominous tumor (*5*, *6*). Herein, we present a case of primary leiomyosarcoma of the abdominal aorta presenting with renovascular hypertension due to tumor embolization in the renal arteries.

Case Report

A 44-year-old previously healthy woman presented at an outside hospital with lumbar back pain and hypertension of 1 month duration. An abdominal CT scan showed bilateral renal infarction; hence, the patient was referred to our hospital for further evaluation. On admission, the blood pressure was 182/94 mmHg and the pulse was 94 bpm. A vascular murmur was audible on the upper abdomen. No edema was noted and the peripheral pulses were all intact. A laboratory workup revealed the data presented in Table 1. Plasma renin activity and aldosterone level were high 16.9 ng/ml/h (normal: 0.2–3.9 ng/ml/h) and 324 pg/ml (normal: 56.9–150.3 pg/ml), respectively. Urinary noradrenalin and dopamine were also high 238 μg/day (normal: <120 μg/day) and 925 μg/day (normal: <925 μg/day), respectively, whereas urinary adrenalin was within the normal range 3 μg/day (normal: <15 μg/day). A repeat abdominal CT (Fig. 1) revealed extensive bilateral renal infarction with a left adrenal mass that had become much greater in size as compared to the CT taken a month earlier. Owing to the rapid growth of the mass, a diagnosis of retroperitoneal hematoma, presumably resulting from an adrenal infarction, was suspected. A post-contrast three-dimensional CT of the aorta (Fig. 2) depicted some irregularity of the wall of the aorta proximal to the origin of the renal

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Table 1. Laboratory Data on Admission

Peripheral blood			Urinalysis		
WBC	9,610	/mm^3	Specific gravity	1.012	
RBC	397	$\times 10^4$/mm^3	Protein	(−)	
Hb	12.3	g/dl	Sugar	(−)	
Ht	35.9	%	Occult blood	(−)	
Plt	31.8	$\times 10^4$/mm^3	Sediment	none	
Na	139	mEq/l	Endocrinology and serology		
K	3.7	mEq/l	PRA	16.9	ng/ml/h
Cl	103	mEq/l			(0.2–2.7 ng/ml/h)
Ca	9.0	mg/dl	PAC	324	pg/ml
IP	3.0	mg/dl			(36–240 pg/ml)
Blood chemistry			Arterial blood gas		
TP	6.8	g/dl	pH	7.433	
Alb	3.7	g/dl	$PaCO_2$	41.2	mmHg
BUN	11	mg/dl/l	PaO_2	89.4	mmHg
Cre	3.6	mg/dl	HCO_3^-	27.1	mmol/l
UA	3.7	mg/dl	Urinary catecholamine (3-fraction)		
AST	27	IU/l	Adrenalin	3	μg/day
ALT	22	IU/l			(<15 μg/day)
LDH	183	IU/l	Noradrenalin	238	μg/day
ALP	272	IU/l			(<120 μg/day)
ChE	31	IU/l	Dopamine	925	μg/day
CPK	518	IU/l			(<700 μg/day)
T. Bil	0.4	mg/dl	Urinary free cortisol	102	μg/day
TC	75	mg/dl			(26–187 μg/day)
TG	180	mg/dl			
CRP	<0.1	mg/dl			

Ht, hematocrit; Plt, platelets; TP, total protein; Alb, albumin; BUN, blood urea nitrogen; Cre, creatinine; UA, uric acid; AST, aspartate aminotransferase; ALT, alanine aminotransferase; LDH, lactate dehydrogenase; ALP, alkaline phosphatase; ChE, cholinesterase; CPK, creatine phosphokinase; T. Bil, total bilirubin; TC, total cholesterol; TG, triglyceride; CRP, C-reactive protein; PRA, plasma renin activity; PAC, plasma aldosterone concentration.

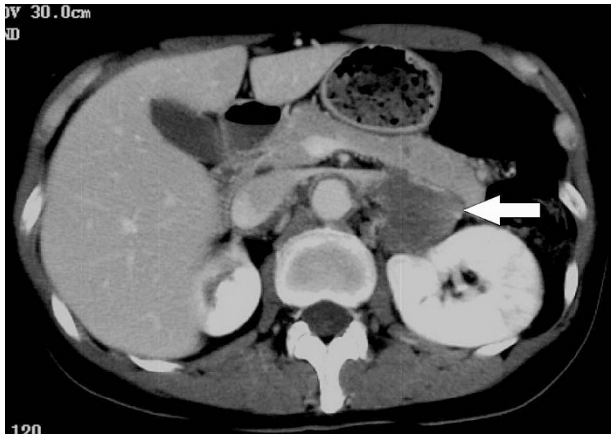

Fig. 1. *Abdominal CT scan image showing bilateral renal infarction and a left adrenal mass (arrow).*

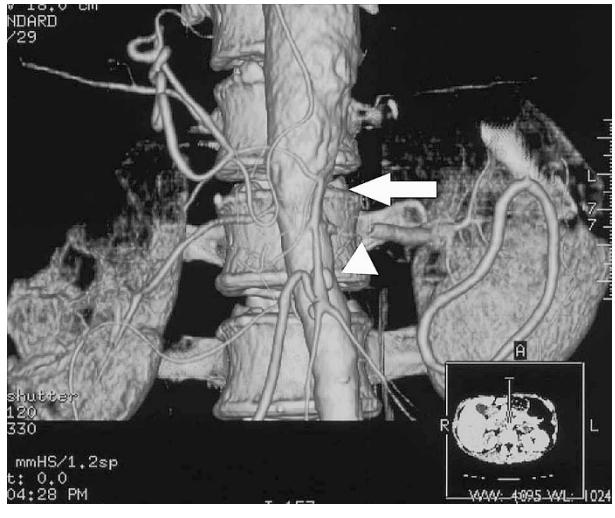

Fig. 2. *Post-contrast three-dimensional CT scan image of the aorta showing some irregularity of the wall (arrow), non-enhanced lesions involving the entire upper pole of both kidneys, and stenosis of the renal arteries (arrowhead), celiac artery and superior mesenteric artery.*

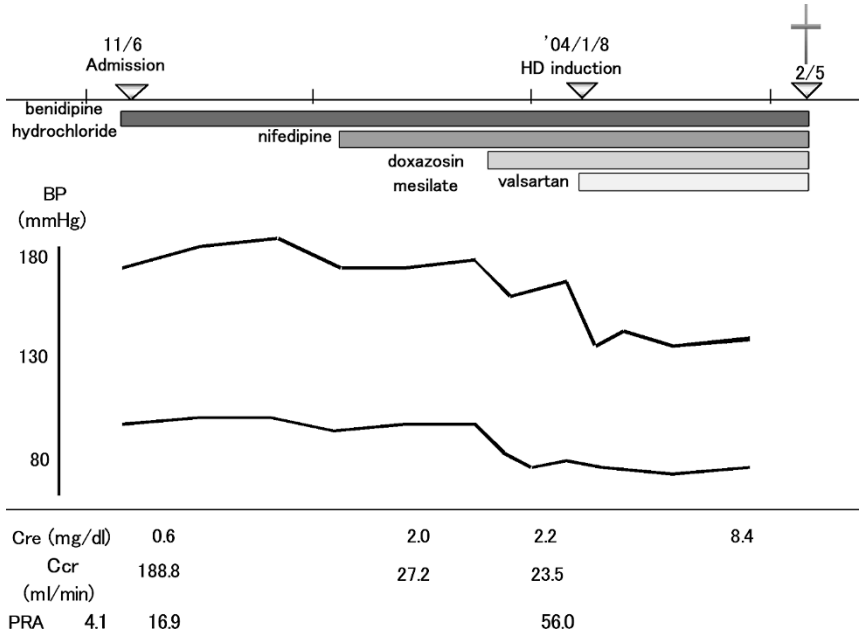

Fig. 3. *Clinical course.*

arteries with an intramural thrombus, non-enhanced lesions involving the entire upper pole of both kidneys, and stenosis of both renal arteries, the celiac artery (CA) and the superior mesenteric artery (SMA). At that time, the diagnosis of Takayasu's disease was suspected but malignancy of the adrenal gland could not be excluded. Consequently, rescue therapy with pulse methylprednisolone (500 mg i.v. daily for 3 days) and heparin sodium (500 units/h i.v.) was started but the patient's renal function rapidly deteriorated and signs of congestive heart failure appeared. An echocardiogram showed a dilated left ventricle with hypokinesia of the left ventricular wall and poor ejection fraction. However, coronary angiography revealed normal coronary arteries. T_1-weighted MRI of the abdomen showed an enlarged ($57 \times 44 \times 70$ mm) hypointense left adrenal mass, which was depicted on T_2-weighted images as area of mixed-signal intensity with a surrounding well-enhanced capsular structure. These findings were compatible with an organized hematoma rather than a lipid-rich adrenal tumor. As the hope of rescuing the renal function faded, we considered giving the priority to the symptomatic treatment. The blood pressure was 170/100 mmHg; we therefore added valsartan, an angiotensin II receptor antagonist, at 80 mg/day to the previously prescribed antihypertensive treatment (benidipine hydrochloride 5 mg/kg body weight, nifedipine 40 mg/day, furosemide 60 mg/day and doxazosin mesilate 8 mg/day). The renal function continued to deteriorate and regular hemodialysis had to be started 3 months after her initial presentation (Fig. 3). A follow-up CT scan (Fig. 4) showed a huge retroperitoneal mass encasing the aorta and invading the pancreatic tail. Therefore, a diagnosis of either primary or secondary adrenal tumor was made. A few days later, the patient developed severe abdominal pain and died with refractory hypotension and severe metabolic acidosis.

The autopsy revealed a tumor in the aorta (Fig. 5) with tumor thrombosis in the CA, SMA and both renal arteries. The adventitial surface of the aorta and these arteries were intact. The left kidney showed partial atrophy due to ischemic change. The tumor had disseminated to the left adrenal gland, pancreas, left kidney, right ovary, lung and peritoneum. The left adrenal gland was filled with partially hemorrhaged tumor tissue and was clearly enlarged. Hemorrhagic infarction of the ileum and ascending colon was observed due to SMA occlusion by the tumor embolism. Pathological examination of the tumor tissue from the left adrenal grand revealed a poorly differentiated malignant neoplasm (Fig. 6). Immunohistological staining revealed that the tumor was positive, with vimentin and α-smooth muscle actin antibodies confirming the smooth muscle origin of the tumor cells (Fig. 7).

Discussion

Retroperitoneal leiomyosarcoma is the second most common retroperitoneal sarcoma after liposarcoma. Three growth patterns have been described: completely extravascular (62%), completely intravascular (5%), and extra- and intravascular (33%) (*7*). Vascular involvement is predominantly venous, whereas leiomyosarcoma of the arterial origin is extremely rare. We have described a case of leiomyosarcoma with extra- and intravascular growth involving the abdominal aorta, in which a definite diagnosis was made at postmortem examination.

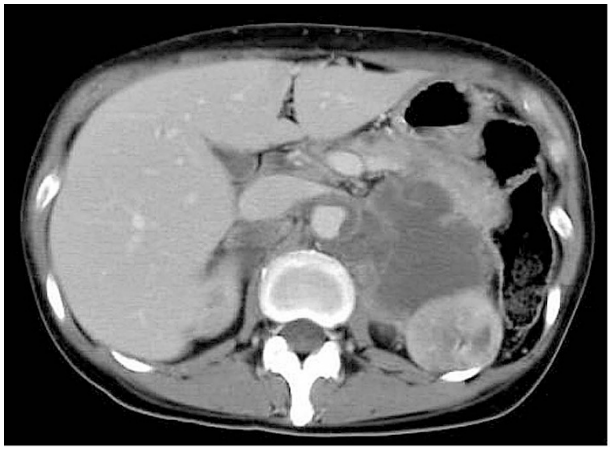

Fig. 4. *Follow-up CT scan image showing a huge retroperitoneal tumor mass encasing the aorta and invading the tail of the pancreas.*

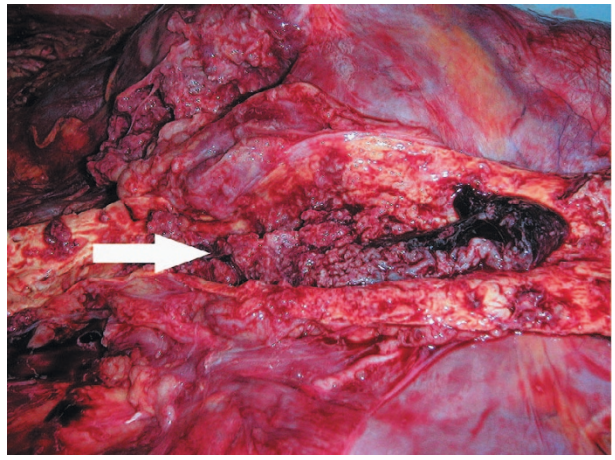

Fig. 5. *Postmortem photograph; a longitudinal section through the aorta reveals fleshy nodular mass lesions attached to its interior surface (arrow).*

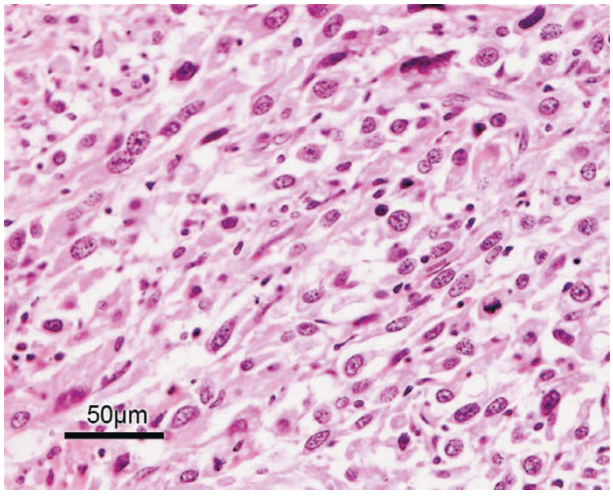

Fig. 6. *Histology of the tumor showing a poorly differentiated malignant neoplasm. A variable proportion of spindle-shaped, round and pleomorphic cells can be seen. Many nuclei appear abnormally large, and an increase in cells undergoing mitosis is seen (hematoxylin and eosin).*

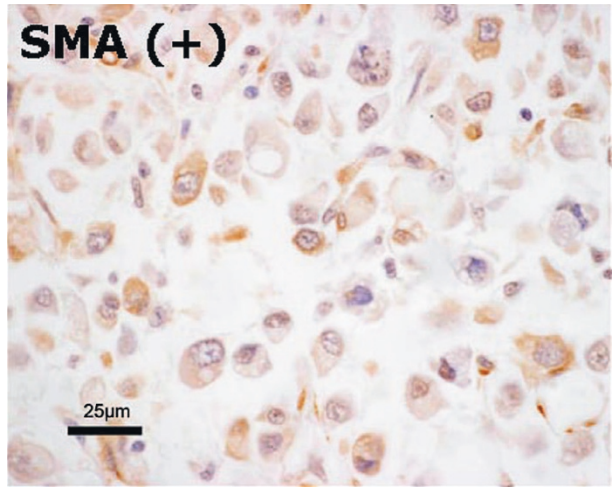

Fig. 7. *Immunohistological staining showing that the tumor was positive with α-smooth muscle actin antibodies, indicating that the tumor cells originated from smooth muscle.*

The big question concerning the present case is whether the tumor originated from the aorta or from the retroperitoneum, because the initial CT scans showed a sizable mass in the retroperitoneal space. Our patient presented with renovascular hypertension due to bilateral renal artery stenosis that was severe enough to cause bilateral renal infarction, and this stenosis was unlikely to have occurred simply due to an outside pressure on the renal arteries, particularly since outside pressure would likely be unilateral. Moreover, the finding of obstructing lesions in the aorta and in multiple visceral arteries on the post-contrast CT suggests the presence of a primary malignant tumor of the aorta with thromboembolic metastasis. Retroperitoneal leiomyosarcoma may invade neighboring blood vessels, including the aorta, often causing aortic rupture (*8*); however, no direct tumor invasion into the aorta was noted at the postmortem examination. Therefore, the origin of this tumor was considered to be the aorta, and the lack of continuity with the aortic tumor indicated a hematogenous metastasis into the retroperitoneal region.

Aortic tumors are classified into two categories according to the site of occurrence in the aortic wall: the intimal-type tumors and the mural-type tumors (*6*, *9*). This case belongs to the former group. Intimal-type tumors grow in the direction of the aortic lumen, extending along the intima or growing as polypoidal masses. They have a high potential for tumor thromboembolization (*10*, *11*). The clinical presentation is

caused by the level of obstruction of the lumen. Our patient presented with severe hypertension and rapid deterioration of renal function due to occlusion of the renal arteries, and ultimately died with ischemic colitis due to occlusion of the SMA. Intimal tumors have dismal prognosis, since the intimal origin is a source of widespread metastasis, and seeding of the tumor to distant sites occurs early. They appear histologically as poorly differentiated mesenchymal tumors, showing immunohistochemical characteristics analogous to those of tumors originating in the myofibroblasts (*12*). Despite advances in aortic imaging techniques, most intimal tumors are initially overlooked, because they are often very similar in presentation to mural thrombi, as was the case in our patient. The initial differential diagnosis also included Takayasu's arteritis, an uncommon disease affecting mainly young women in South East Asia, which is characterized by chronic inflammation and occlusion of the aorta and its major branches (*13*). However, when the first follow-up CT scan showed a rapidly growing retroperitoneal mass, hematoma was suspected and thus no biopsy was done, and when the diagnosis was finally made, the tumor had already reached an advanced stage and the patient died soon afterward.

Chronic or slowly progressing bilateral renal artery stenosis does not cause the renin-angiotensin activation, because of sodium retention due to renal dysfunction. In contrast, acute progression of renal artery stenosis induces obvious activation of the renin-angiotensin system, as in the present case. Increased sympathetic nerve activity has been observed in several renal diseases, including renovascular hypertension (*14*). In our case, the urinary catecholamines (noradrenalin and dopamine) were also increased, indicating an elevation of sympathetic nerve activity. In the clinical course, both valsartan and doxazosin mesilate were effective as antihypertensive agents. Hence, methylprednisolone had little effect on the blood pressure in this case (Fig. 3).

In conclusion, the present case serves to remind physicians to consider the rare possibility of primary malignant tumor of the aorta with tumor embolization into the renal arteries in any patient with renovascular hypertension, particularly in the absence of risk factors for atherosclerosis. The presence of stenotic lesions in other visceral arteries and a dramatic clinical course would further support this possibility. Radiologists should also be aware of the existence of such tumors, and keep in mind that behind a retroperitoneal mass there might be hidden a tumor of the aorta.

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