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Case Report

Cardial leiomyosarcoma with multiple lesions involved: a case report

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Abstract: Background: Leiomyosarcoma of the heart is extremely rare and may involve many symptoms. The outcome is poor and the median survival is only 6 months. Case presentation: A 43-years-old female patient complained of palpitation and dyspnea and was diagnosed as bilateral iliac vein-inferior vena cava-right atrium-pulmonary masses. Pathological diagnosis was leiomyosarcoma of vascular origin and she survived for 12 months after surgery. Conclusion: According to the summarization of 30 vascular leiomyosarcoma cases with heart involved we can find that surgical resection is the basic treatment for cardiac leiomyosarcoma, and surgery combined with chemotherapy may be able to further prolong survival.

Keywords: Leiomyosarcoma, cardiac tumor, vascular origin, therapy

Background

Leiomyosarcoma is a malignant mesenchymal tumor originated from the intestinal wall smooth muscle and intestinal smooth muscle vascular or intestinal mucosa muscle, occupied 5% to 10% of all soft tissue tumors. Leiomyosarcoma of vascular origin often occurs in inferior vena cava [1]. Primary cardiac tumor is extremely rare, and its incidence is only 0.001% to 0.03% [2], so cardiac leiomyosarcoma is almost metastasis or infiltration from adjacent organ [3]. Leiomyosarcoma of cardiac may involve many symptoms, such as dyspnea, chest pain, arrhythmia, heart failure and so on. The treatment outcome for patients remains poor [4], and the median survival after diagnosis was 6 months [5].

Case presentation

A 43-years-old female patient went to our hospital with a complaint of “palpitation and dyspnea for 1 year, aggravated for 1 month”. Additional echo was observed in right atrium by cardiac ultrasound in community hospital, accompany with tricuspid valve obstruction and pulmonary hypertension. Five years ago she was diagnosed as uterine leiomyosarcoma in another hospital and hysterectomy was given, without radiation and chemotherapy after surgery. Systolic murmurs were audible in tricuspid valve area.

Routine biochemical test (hepatic function, renal function, myocardial calcium protein) were normal, hemoglobin is 9 g/L. No abnormal was observed in thoracic-abdominal aorta CT. But venous system: low density filling defect areas were detected in right atrium, inferior vena cava, bilateral iliac vein, double main pulmonary artery and its branches (**Figure 1**). Abdominal and pelvic cavity enhanced CT did not find space occupying lesion. According to ultrasound and CT, we diagnosed her as bilateral iliac vein-inferior vena cava-right atrium-pulmonary masses. Cardiac surgery and general surgery were combined together for the operation. After separating right iliac artery and iliac vein, vena cava, we incised inferior vena cava and the masses were gray funicular, adhered with and almost completely blocked the inferior vena cava. After incised right atrium, we saw the masses growing from the inferior vena cava

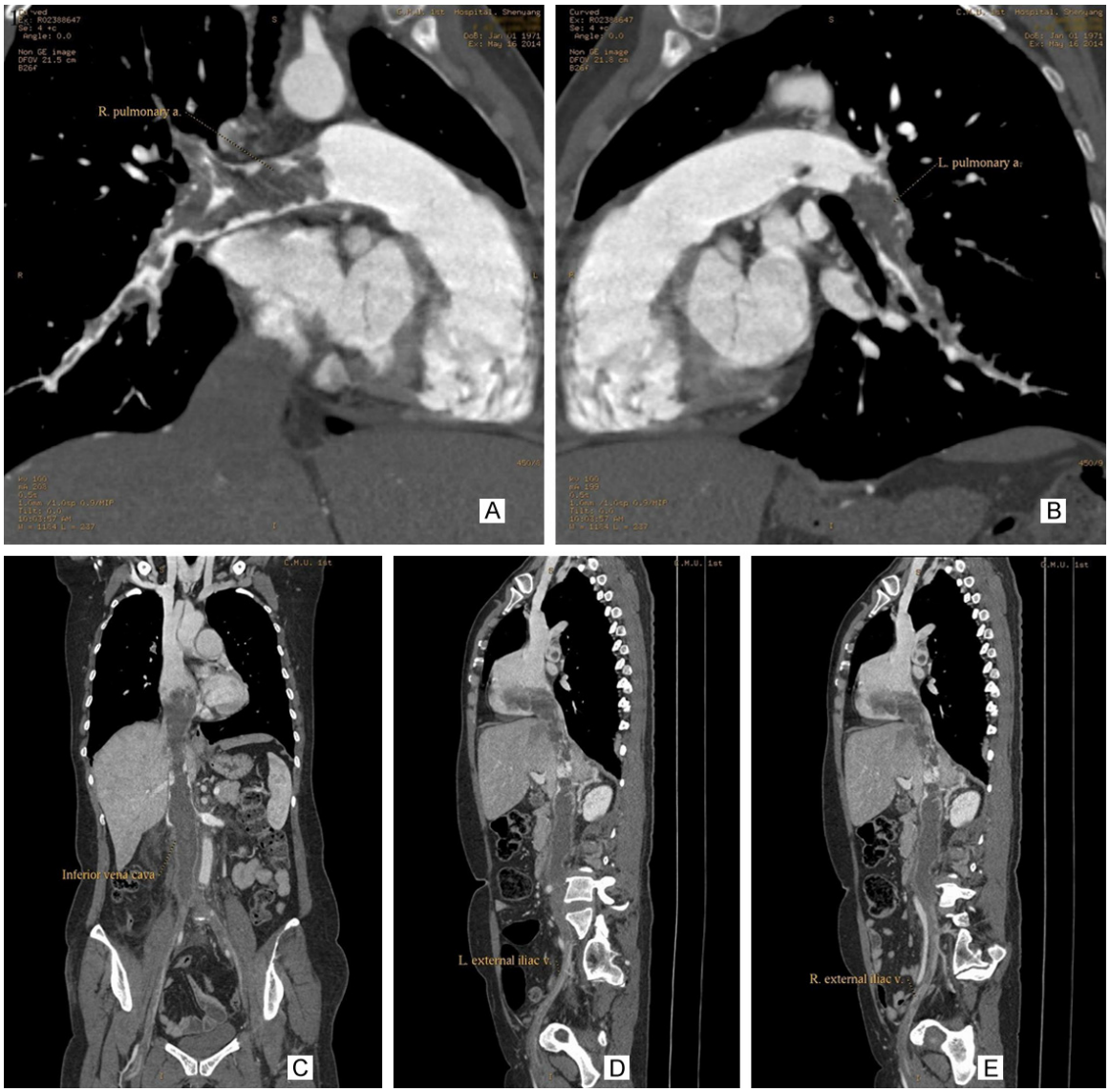

Figure 1. Computed tomography. A. Right pulmonary artery and right atrium; B. Left pulmonary artery; C. Inferior vena cava; D. Left iliac vein; E. Right iliac vein. Low density filling defect areas were detected in the above parts.

to right atrium, obstructing tricuspid valve, we dissected the neoplasm completely. Finally we incised left and right pulmonary artery, removed the masses completely under extracorporeal circulation. Ventilator and ECMO technique were used after operation, finally recovered. Pathological diagnosis was leiomyosarcoma of vascular origin (**Figure 2**). She did not receive radiation and chemotherapy, and no signs of recurrence 12 months after operation.

Conclusion

We reported a case of vascular leiomyosarcoma involving the iliac vein, inferior vena cava, right atrium and pulmonary artery. Montemezzi et al [6] firstly reported the case of uterine leiomyosarcoma with heart involved in 1966. 30 vascular leiomyosarcoma cases with heart involved were summarized published on PubMed in recently 20 years (**Table 1**). In these cases, the average age of onset was 53.6 ± 15.3, and 25 cases were between 40-70 years old. There were many symptoms including dyspnea, chest and back pain, cough, swelling, dizziness, fever, and cardiac arrhythmias, dyspnea is the most common symptom, accounting for about 2/3. In the 20 female patients, four patients had history of uterine leiomyosarco-

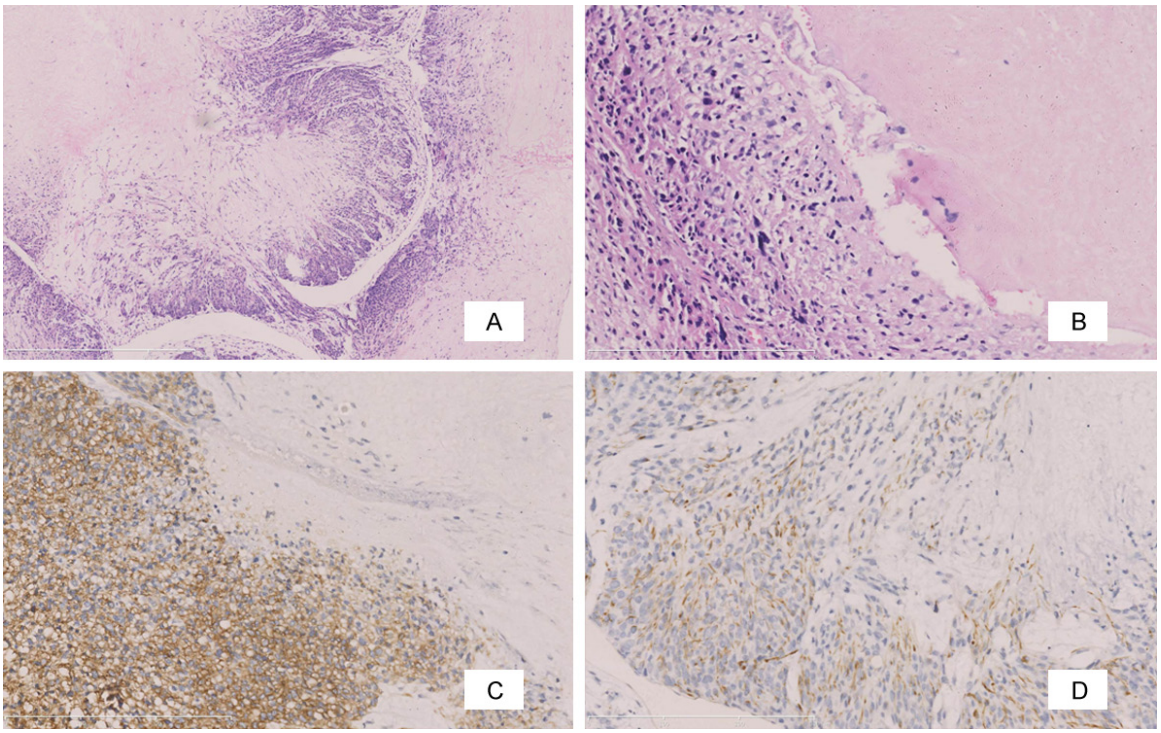

Figure 2. Pathology. A. Hematoxylin-eosin (HE) stains show that vascular fissure can be seen in the tumor organization, short spindle cells distribute as plate and arrange densely, accompanied by obvious necrosis (50×); B. HE stains show that tumor cell atypia, nuclear hyperchromatism, different sizes are obvious, tumor giant cells and mitosis are visible (>5/10HPF) (200×); C, D. Immunohistochemical show that myogenic markers Actin (SMA) and Desmin are expressed by the tumor cells (200×).

ma; two had hysterectomy for menorrhagia, one suffered from the iliac vein leiomyosarcoma, one suffered from breast cancer, one suffered from uterine fibroids. 16 cases were only heart involved, 14 were multi-part involved, the most common cardiac involvement was right atrium, accounting for 14 cases, followed by left atrium (7), right ventricular (6), pulmonary artery (6), left ventricular (2), the most rare is the coronary and pulmonary vein, 1 case respectively. 25 cases received surgical treatment, and after operation, 1 received radiotherapy, 10 received chemotherapy alone, 1 received both. The shortest survival was seven days, the longest was 36 months. From the data, we found that female accounted for the vast majority of this disease, and some female patients had a history of uterine disease, which suggesting that physiological characteristics of female may be closely related to this disease. In addition, the location and extent of lesions involving significantly affect the prognosis. The mean follow-up survival time of single cardiac cavity involved patients was 15 months, more than one was 9.5 months, and multi locations and organ was six months, only two months in the patients with other organ involved, four patients died and three of which suffered from other organ involved, and four patients appeared recurrence or metastasis. Surgical resection is the basic treatment [7], patients who underwent a complete resection show a better survival compared with patients who did not [8]. According the **Table 1** we can find that the mean survival time was 12 months in patients given operation and chemotherapy, and surgery alone was 10.4 months, surgery combined with chemotherapy may be able to further prolong survival. To prevent metastasis and possibility of incomplete tumor excision, chemotherapy can consider after operation [9]. Because of low sensitivity and possibly damage to myocardium and cardiac, effect of radiotherapy is limited. Reports suggest combination of radiotherapy and chemotherapy after surgery can improve survival compared with radiotherapy only [10]. Therefore, because of its rare frequency, more cases and longer follow-up are

Table 1. Summary of 30 cases of leiomyosarcoma with heart involvement

	Age	Sex	Symptom	History	Lesions	Operation	Radiotherapy	Chemotherapy	Size of tumor (cm)	Recurrence/Metastasis/Death	Follow-up
1	65	F	Dyspnea	N	Uterine, IVC, RA, PA	Y	N	Y	7*2*2	N	14 m
2	57	F	Abdominal pain	Uterine leiomyoma	IVC, RA	N	N	N	NA	NA	NA
3	67	F	Dyspnea, nausea	Breast cancer	LV, Coronary	Y	N	N	1*1*1	D	7 d
4	55	F	Dyspnea	Uterine leiomyoma	RA	Y	N	N	NA	N	16 m
5	26	M	Dyspnea	N	LA	Y	N	N	8.5*6.7*3.6	NA	NA
6	86	F	Dyspnea	N	LA	Y	N	N	NA	N	15 m
7	49	F	Dyspnea	Uterine fibroids	RA, RV, PA	Y	N	N	NA	NA	NA
8	56	F	Dyspnea, fever	N	RA, Uterine	Y	N	N	4*3*4	NA	NA
9	42	F	Abdominal pain	N	IVC, RA, RV	Y	Y	N	6*8*3	N	3 m
10	36	M	Dyspnea	N	PA, RA, RV	Y	N	Y	NA	N	4 m
11	69	F	N	N	LA	Y	N	Y	NA	D	9 m
12	65	M	N	Iliac vein leiomyosarcoma	PA, RA	Y	N	Y	NA	N	6 m
13	33	F	Dyspnea	N	LA	Y	Y	Y	NA	R,M	33 m
14	69	F	Tachycardia	Uterine leiomyoma	RA, Both lungs	N	N	Y	NA	N	1 m
15	45	M	Dyspnea	N	PA, RV	Y	N	Y	9.5*5*4	NA	NA
16	29	F	N	N	LA	Y	N	N	NA	N	8 m
17	49	M	Dizziness	N	LV	Y	N	N	3*2.5*2	N	36 m
18	35	F	Fever, asthenia	N	RA	Y	N	Y	0.8*0.8*0.88	NA	NA
19	58	F	Syncope	N	RV	Y	N	N	NA	NA	NA
20	64	F	Chest pain and dyspnea	N	PA	Y	N	Y	NA	N	25 m
21	22	M	Dyspnea	N	PV, LA, Brain	Y	N	N	6.4*5*4	M,D	6 m
22	54	M	Abdominal pain	N	IVC, RA	Y	N	N	NA	N	12 m
23	73	M	Dyspnea	N	LA	Y	N	N	NA	N	3 m
24	45	M	Dyspnea, chest pain	N	RA, RV	Y	N	Y	NA	M	24 m
25	70	F	Dyspnea	Hysterectomy for hemorrhage	RV	Y	N	N	4*2.3*2	N	7 m
26	50	F	Edema, ascites	N	RA	N	N	Y	NA	N	6 m
27	54	F	Cough, dyspnea	Hysterectomy for menorrhage	PV	Y	N	N	3.5*2.5*2.5	NA	NA
28	53	F	Backache, arthralgia	N	RA	N	N	N	NA	N	21 m
29	61	F	Right flank pain	N	RA, IVC, Right renal vein	Y	N	N	NA	D	1 m
30	70	M	Dyspnea	Uterine leiomyoma	LA	N	N	N	NA	M	3 m

RA = Right atrium, RV = Right ventricle, LA = Left atrium, LV = Left ventricle, PV = Pulmonary vein, PA = Pulmonary artery, IVC = Inferior vena cava, N = None, NA = Not Applicable, N = No, R = Recurrence, M = Metastasis, D = Death.

required to assess more effective therapy other than surgical resection.

Disclosure of conflict of interest

None.

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