A Rare Case of Pulmonary Leiomyosarcoma

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ABSTRACT

Leiomyosarcoma commonly occurs in the abdomen, retroperitoneum, large blood vessels, and uterus[1]. Cardiac leiomyosarcoma is a rare and highly aggressive sarcoma. We reported a case of a 63-year-old male with pulmonary artery leiomyosarcoma. Transthoracic echocardiography showed a large 4.4×2.3 cm hypoechoic mass in the right ventricular outflow tract and pulmonary artery. Computed tomography pulmonary angiography showed a filling defect in a similar location. The initial impression was PE, but a tumor was not ruled out. An emergency surgery was performed due to progressively worse chest distress and dyspnea. A yellow mass that had adhered to the ventricular septum and pulmonary artery wall was detected to be compressing the pulmonary valve. Immunohistochemistry confirmed tumor cells positive staining for Desmin and smooth muscle actin and negative staining for S-100, CD34, myogenin, or myoglobin, and KI67(+)80%, indicating leiomyosarcoma. Pulmonary leiomyosarcoma showed a side-inserted heart chamber filling defect in CTA and should be excised when the patient suddenly deteriorated.

Key words: *Pulmonary leiomyosarcoma, pulmonary embolism, diagnosis.*

INTRODUCTION

Leiomyosarcoma commonly occurs in the abdomen, retroperitoneum, large blood vessels, and uterus.[1] Cardiac leiomyosarcoma is a rare and highly aggressive sarcoma. It accounts for 1% of all cardiac tumors and 8-9% of all cardiac sarcomas.[2] Cardiac leiomyosarcoma mostly involves the left atrium, rarely occurring in the pulmonary artery. Pulmonary artery leiomyosarcoma has been commonly misdiagnosed as pulmonary embolism in some reports.[3-5] This paper reported a case that manifested as pulmonary artery leiomyosarcoma that mimicked a pulmonary embolism.

CASE ILLUSTRATION

A 63-year-old male was admitted to our hospital with chest distress and dyspnea. One year before his admission, chest distress initially began and progressively escalated. Then, dyspnea subsequently emerged 2 months prior to admission. When he was admitted, his physical indicators were determined as follows: blood pressure of 120/80 mmHg, pulse of 93 bpm, respiratory rate of 22 breaths per minute, body temperature of 36.7℃, and SPO2 92%. Physical examination revealed a systolic murmur. Transthoracic echocardiography (TTE) showed a large 4.4×2.3 cm hypoechoic mass in the right ventricular outflow tract and pulmonary arterial trunk and enlarged right ventricle. Color doppler

flow imaging (CDFI) showed a filling defect in the pulmonary artery, high systolic peak velocity (4.52 m/s) (**Figure 1A**) and moderate tricuspid regurgitation. Pulmonary artery pressure was approximately 128 mmHg.

For a definite diagnosis, the patient was thoroughly examined. Computed tomography pulmonary angiography (CTA) showed a filling defect from the right ventricular outflow tract to the pulmonary trunk (**Figures 1B, 1C,** and **1D**). Chest computed tomography (CT) revealed a few small nodules. Abdominal CT or cranial CT showed no lesions, and ECG was sinus rhythm. The initial diagnosis result was pulmonary embolism, but D-dimer level was 300 µg/L. Then, we speculated it could be a tumor. However, CA-125, CA-199, CEA, and AFP were normal. Thus, we guessed that it could be a myxoma, a common cardiac tumor. Unfortunately, his dyspnea had been progressively worsening, so we performed an emergency operation for the patient to dredge the occlusion.

The operation was performed on the cardiopulmonary bypass. When the right ventricular outflow tract and pulmonary artery were opened longitudinally, a yellow mass was detected on the ventricular septum and pulmonary artery wall (**Figures 2E** and **2F**). It almost occupied the entire right ventricular outflow tract and pulmonary artery trunk, compressing pulmonary valve. The tumor was excised substantially and then a tricuspid annuloplasty ring was inserted. TEE showed mild pulmonary valve regurgitation. A histological examination of the resected specimen showed proliferation of spindle cells with hyperchromatic or pleomorphic nuclei (**Figure 2G**). Immunohistochemistry confirmed the tumor cells positive staining for Desmin and smooth muscle actin and negative staining for S-100, CD34, Myogenin, or S-100 protein, indicating leiomyosarcoma.

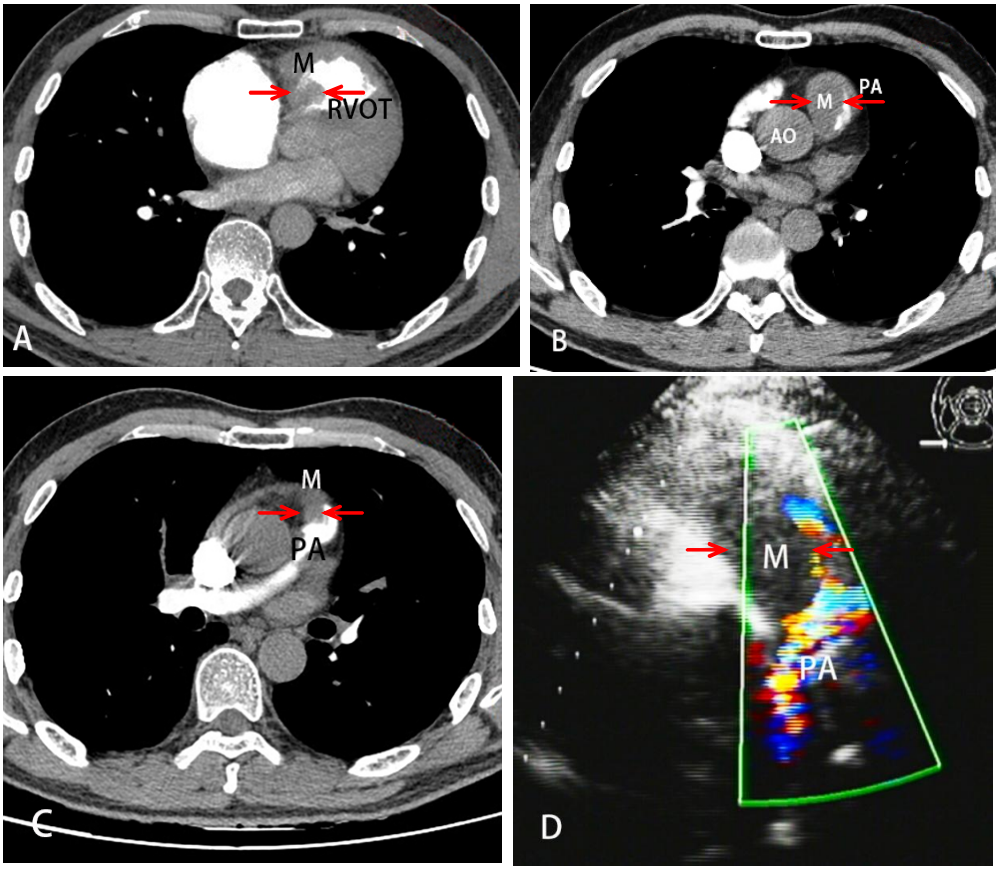

Figure 1. A) Pulmonary computed tomography angiography showing intravascular filling defects of pulmonary trunk occupying right ventricular outflow tract. B) The mass nearly filling the entire pulmonary cavity. C) The tumor extending distal to the pulmonary artery branch. D) Transthoracic echocardiography showing a filling defect of the pulmonary artery.

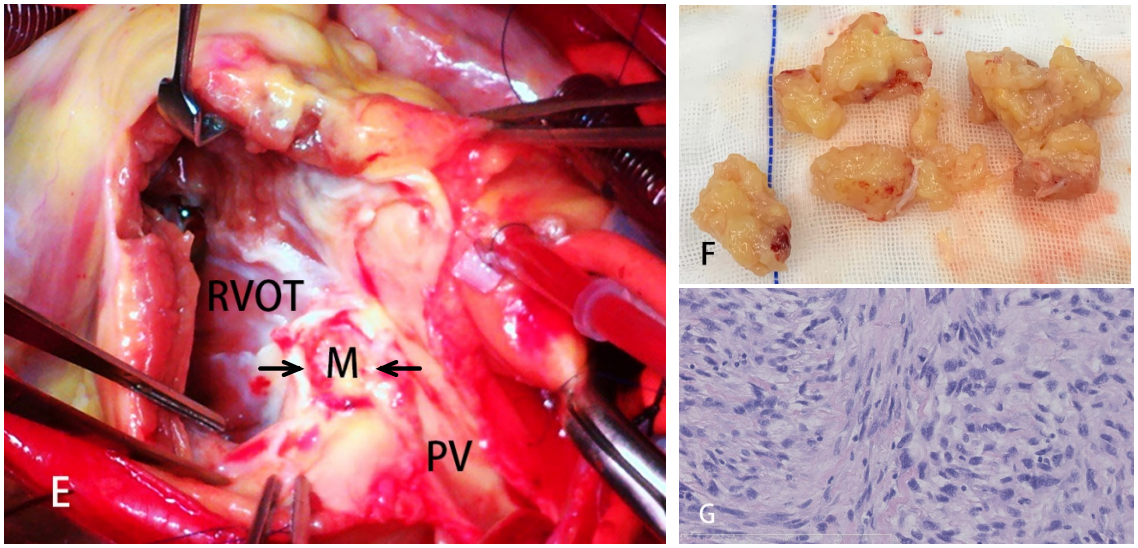

Figure 2. E) The tumor adhering to the right ventricular outflow tract and pulmonary artery. F) Leiomyosarcoma presenting as a yellow mass. G) Tumor composed of interlacing fascicles of pleomorphic spindle cells with abundant mitotic figures (H&E, high power).

DISCUSSION

Cardiac leiomyosarcoma is a rare tumor of the cardiovascular system, accounting for 1% of all cardiac tumors and 8-9% of all cardiac sarcomas.[2] Primary cardiac leiomyosarcoma primarily occurs in the left atrium, and other locations, including the right ventricle, the right atrium, and the left ventricle.[2] Pulmonary arteries are rarely involved.

If a filling defect within a pulmonary artery indicates a PE,[6] is it really pulmonary embolism? Although the first diagnosis result was PE, we should consider it could be a tumor. Some researchers reported that the patients with pulmonary arterial leiomyosarcoma were diagnosed as pulmonary embolism pre-operation, but their symptoms after anticoagulant therapy did not improve and even worsened gradually, even leading to death.[3,4] We should consider the possibility of pulmonary leiomyosarcoma when discovering pulmonary artery filling defect. In addition, we should excise the mass without hesitation when symptoms were worsened.

It is difficult to differentiate PE from tumors. Pulmonary CTA and ventilator-perfusion are essential for diagnosing pulmonary embolism.[6] Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) has also been described as an effective tool for diagnosing intraluminal malignant neoplasms in certain cases,[5,8,9] but the technique is seldom administered to patients duo to its major bleeding risk, particularly in patients with pulmonary hypertension. From the patient CTPA, we found that the filling defects were located on one side of the pulmonary atrial and protruded into the cavity or the opposite side. In addition, the patient with pulmonary embolism in the CTPA showed a double-track sign. The thrombus usually adheres to atrial walls in a ring shape, whereas the leiomyosarcoma manifests invasion and bulge from the vessel lumen. Thus, we believed that a side filling defect inserted into the heart chamber could be a tumor and should be excised when symptoms suddenly deteriorated.

Surgery is applicable for patients with pulmonary leiomyosarcoma due to rare initial metastasis.[10] The average survival for patients without surgery is only 6 months and surgery can extend it to 24 months.[11] However, the local recurrence rate is relatively high[10] and adjuvant therapy should be considered.[12] The efficacy of adjuvant chemotherapy also remains controversial.[13] Some scholars reported that neoadjuvant chemotherapy could shrink the size and edges of cardiac sarcoma, thus providing an opportunity for complete resection.[14] Therefore, it is extremely important to consider the possibility of pulmonary artery leiomyosarcoma if radiologic examinations demonstrate pulmonary filling defects.

CONCLUSION

Pulmonary leiomyosarcoma is rare and neglected easily. We believed that pulmonary leiomyosarcoma could be manifestations that CTA showed a side-inserted heart chamber filling defect and should be excised when the patient suddenly deteriorated.

CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

ETHICS STATEMENT

Informed consent was obtained from the patient to publish this case report.

REFERENCES

1. Mangla A, Yadav U. Leiomyosarcoma. In: StatPearls, editor. Cancer, Treasure Island (FL): StatPearls Publishing; 2020.
2. Li X, Chen Y, Liu J, et al. Cardiac magnetic resonance imaging of primary cardiac tumors. Quant Imaging Med Surg. 2020;10(1): 294-313.
3. Muganlinskaya N, Guzman A, Dahagam C, Selinger SR. When a pulmonary embolism is not a pulmonary embolism: a rare case of primary pulmonary leiomyosarcoma. J Community Hosp Intern Med Perspect. 2015;5(6):29624.
4. Flaherty G, McCarthy P, Mortimer G. Pulmonary artery leiomyosarcoma: an unusual cause of shortness of breath. Ir J Med Sci. 2011;180(1):275-8.
5. Romero Francés L, Royo Prats JA. Pulmonary artery leiomyosarcoma diagnosed by magnetic resonance, PET-CT and EBUS-TBNA. Leiomiosarcoma de arteria pulmonar, diagnóstico mediante resonancia magnética, PET-TC y EBUS-TBNA. Arch Bronconeumol. 2017;53(9):522-3.
6. Kruger PC, Eikelboom JW, Douketis JD, Hankey GJ. Pulmonary embolism: update on diagnosis and management. Med J Aust. 2019;211(2):82-7.
7. McCabe BE, Veselis CA, Goykhman I, Hochhold J, Eisenberg D, Son H. Beyond pulmonary embolism; nonthrombotic pulmonary embolism as diagnostic challenges. Curr Probl Diagn Radiol. 2019;48(4):387-92.
8. Al-Saffar F, Ibrahim S, Seeram V, Bajwa AA, Shujaat A. Use of endobronchial ultrasound to evaluate nonthrombotic endovascular lesions in pulmonary arteries: A systematic review. J Bronchol Interv Pulmonol 2015;22(1):28-32.
9. Caraway NP, Salina D, Deavers MT, Morice R, Landon G. Pulmonary artery intimal sarcoma diagnosed using endobronchial ultrasound-guided transbronchial needle aspiration. Cyto Journal 2015;12:3.
10. Vakilian F, Shabestari MM, Poorzand H, Teshnizi MA, Allahyari A, Memar B. Primary pulmonary valve leiomyosarcoma in a 35-year-old woman. Tex Heart Inst J. 2016;43(1):84-7.
11. Schoen FJ, Mitchell RN. The heart. In: Kumar V, Abbas AK, Aster JC, editors. Robbins and Cotran pathologic basis of disease. 9th ed. Philadelphia: Elsevier Saunders; 2015. p. 523-78.
12. Abu Saleh WK, Ramlawi B, Shapira OM, et al. Improved outcomes with the evolution of a neoadjuvant chemotherapy approach to right heart sarcoma. Ann Thorac Surg. 2017;104(1):90-6.
13. Ratan R, Patel SR. Chemotherapy for soft tissue sarcoma. Cancer 2016;122:2952–60.
14. Tanaka K, Iwata T, Nishii K, et al. A case of primary pulmonary leiomyosarcoma completely resected after neoadjuvant chemotherapy. Surg Case Rep. 2019;5(1):91.