What is the filling defects of pulmonary trunk: a rare case of pulmonary leiomyosarcoma

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Abstract

Background: Leiomyosarcoma occurs commonly in the abdomen, retroperitoneum, large blood vessels, and uterus[1]. Cardiac leiomyosarcoma is a rare and highly aggressive sarcoma.

Methods and Results: Here, we report the case of a 63-year-old male with pulmonary artery leiomyosarcoma. Transthoracic echocardiography showed a large 4.4×2.3 cm hypoechoic mass in right ventricular outflow tract and pulmonary artery. Pulmonary computed tomography angiography showed a filling defect in similar location. Surgery was performed due to tend to occlusion. A yellow mass adhered to ventricular septum and pulmonary artery wall was detected and compressed pulmonary valve. By immunohistochemistry, the tumor cells stained positive for Desmin and smooth muscle

actin, and stained negative for S-100, CD34, myogenin, or myoglobin, KI67(+)80%, supported leiomyosarcoma.

Conclusion: The patient had recovered from surgery and had been on following-up.

Key words: Pulmonary leiomyosarcoma, Pulmonary embolism, Diagnosis

1 | Introduction

Leiomyosarcoma occurs commonly 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 left atrium, rarely occurs in pulmonary artery. Pulmonary artery leiomyosarcoma was commonly misdiagnosed as pulmonary embolism in some reports[3][4][5]. Here, we report a case that pulmonary artery leiomyosarcoma mimicked pulmonary embolism.

2 | Case report

A 63-year-old male was admitted to our hospital with chest distress and dyspnea. One years before his admission, chest distress was presented and progressively escalated. Then, dyspnea subsequently emerged 2 months prior to admission. When he was admission, his blood pressure was 120/80 mm Hg, pulse 93 bpm, respiratory rate 18 breaths per minute, temperature 36.7℃, SPO_2 96%. Physical examination revealed an ejection 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 ventricular. CDFI showed a filling defect of pulmonary artery, high systolic peak velocity (4.52 m/s) (Fig.1A) and moderate tricuspid regurgitation. Pulmonary artery pressure was approximately 128 mmHg.

The first impression was pulmonary embolism, but the patient was no risk factors of thrombosis and D-dimer levels were 300ug/L. To diagnosis definitely, The patient was

thoroughly investigated. Pulmonary computed tomography angiography (CTA) showed a filling defect was from the right ventricular outflow tract to pulmonary trunk (Fig.1 B, C, D). Chest computed tomography (CT) revealed a few small nodules. Abdominal CT and cranial CT showed no lesions. ECG was sinus rhythm. Thus We considered the filling defect was a tumor. Then, we performed surgery for the patient to dredge occlusion.

Surgery was performed on cardiopulmonary bypass. When right ventricular outflow tract and pulmonary artery was opened longitudinally, a yellow mass located on ventricular septum and pulmonary artery wall was detected (Fig.2 E, F). It almost occupied the entire right ventricular outflow tract and pulmonary artery trunk, compressed pulmonary valve. The tumor was excised substantially and tricuspid annuloplasty ring was inserted. TEE showed mild pulmonary valve regurgitation. Histological examination of the resected specimen showed proliferation of spindle cells with hyperchromatic or pleomorphic nuclei (Fig.2 G). By immunohistochemistry, the tumor cells stained positive for Desmin and smooth muscle actin, and stained negative for S-100, CD34, Myogenin, or S-100 protein, supported leiomyosarcoma.

3 | Discussion

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

Pulmonary arterial mass are common considered as pulmonary embolism or bacterial vegetations. Pulmonary valve bacterial vegetations often attach to the pulmonary valve and may form thrombosis on the surface, even larger mass on ultrasound, causing occlusion. Intermittent fever and positive blood culture conduce to distinguish the mass. Pulmonary arterial leiomyosarcoma usually shows a hypoechoic, irregular, large mass on TTE and pulmonary CTA shows a filling defects of pulmonary artery. When it causes pulmonary artery occlusion, subsequently resulting in an enlarged right atrioventricular, tricuspid regurgitation, and pulmonary arterial hypertension, it is

difficult to differentiate from pulmonary embolism. Some literatures reported that the patients with pulmonary arterial leiomyosarcoma were diagnosed as pulmonary embolism pre-operation, given in anticoagulant therapy, but their symptoms didn't still relieve and gradually worse, even leading to death[3][4]. Although pulmonary CTA and ventilator-perfusion are essential for the diagnosis of pulmonary embolism[6], it is still difficult to identify them. When imaging signs specific for tumor embolism include dilated and beaded peripheral arteries, intravenous injection contrast enhancement of intravascular filling defects[7], the tumor is highly suspected. D-dimer levels helps to exclude pulmonary embolism according to the case. It was difficult to distinguish benign and malignancy. Myxomas was commonly and originated from left atrium, rarely located pulmonary artery or right ventricular outflow tract. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) has also been described as an effective tool to diagnose intraluminal malignant neoplasms in certain cases[5][8][9]. But the technique is seldom used in patients duo to the major risk of its bleeding, particularly in patients with pulmonary hypertension. Due to its occlusion and prolong his life temporarily, we performed surgery.

Surgery was practicable for patients with pulmonary leiomyosarcoma due to rarely initial metastasis[10]. The average survival of patients without surgery is only 6 months, and surgery can be extended to 24 months[11]. But local recurrence rate is relatively high[10], the adjuvant therapy should be considered[12]. The efficacy of adjuvant chemotherapy also remains controversial[13]. At present, some scholars reported that neoadjuvant chemotherapy could shrink the size and edge of cardiac sarcoma, providing opportunity for complete resection[14]. Therefore, it is critically important to consider the possibility of pulmonary artery leiomyosarcoma if radiologic examinations demonstrate pulmonary filling defects.

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.

Reference

[1] Mangla A, Yadav U. Leiomyosarcoma. In: StatPearls, editor. Cancer, Treasure Island (FL): StatPearls Publishing; 2020.

[2] Li X, Chen Y, Liu J, Xu L, Li Y, Liu D, et al. Cardiac magnetic resonance imaging of primary cardiac tumors[J]. 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):275-278.

[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):522-523.

[6] Kruger PC, Eikelboom JW, Douketis JD, Hankey GJ. Pulmonary embolism: update on diagnosis and management. Med J Aust. 2019;211(2):82-87.

[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):387-392.

[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. Cytojournal 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):84-87.

[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, Al Jabbari O, Ravi V, Benjamin B, et al. Improved Outcomes With the Evolution of a Neoadjuvant Chemotherapy Approach to Right Heart Sarcoma. Ann Thorac Surg. 2017;104(1):90-96.

[13] Ratan R and Patel SR. Chemotherapy for soft tissue sarcoma. Cancer 2016; 122: 2952–2960.

[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.

Legends for Figures

Fig.1 A) Transthoracic echocardiography shows a filling defect of pulmonary artery. B) Pulmonary computed tomography angiography shows intravascular filling defects of pulmonary trunk occupied in right ventricular outflow tract. C) The mass almost fills the entire pulmonary cavity. D) The tumor extends distal to the pulmonary artery branch.

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