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CASE REPORT: LEIMYOSARCOMA OF THE PULMONARY VALVE

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ABSTRACT

Myxomas in the heart are the most common type of benign primary tumor, accounting for over 95% of cases. Malignant tumors such as rhabdomyosarcoma, angiosarcoma, fibrosarcoma or leiomyosarcoma are less common and are usually found in the left heart rather than the right. According to the literature, intimal sarcoma is the most common type, accounting for about 42%. Intimal sarcoma includes all the types mentioned above.We report a very rare case: leiomyosarcoma of the pulmonary artery valve. A 57-year-old male patient was admitted due to fatigue. Ultrasound and MSCT showed images suggestive of a myxoma in the pulmonary artery trunk and left pulmonary artery. Abdominal MSCT and coronary angiography yielded normal results. Surgery to remove the tumor en bloc followed by pulmonary valve replacement by tissue valve. The postoperative course was uneventful, without complications, and he was discharged after 7 days. Immuno-histopathology results shoewd a poorly differentiated leiomyosarcoma.

Conclusion: Accurate and early diagnosis of malignant pulmonary artery tumors remains a challenge. MSCT is a useful imaging tool to help determine the cause.

Keywords: *Myxoma in the heart, leiomyosarcoma, intimal sarcoma.*

1. INTRODUCTION

Myxomas in the heart are the most common type of benign tumor. However, malignant myxosarcoma or intimal sarcoma, including angiosarcoma, rhabdomyosarcoma, leimyosarcoma, fibrosarcoma… can still occur, albeit rarely. These forms are more common in the left heart than the right, and in the atrium more than the ventricle [1]. Pulmonary artery sarcoma is very rare.

2. CASE REPORT

Male patient, 57 years old. Reason for admission: fatigue, shortness of breath when lying with head low. Echocardiography showed 39x15mm tumor attached to the pulmonary artery valve, fairly homogeneous density, mobile with the valve leaflet, prolapsed into the right ventricle during diastole, causing mild right ventricular outflow tract obstruction with maximum transvalvular pressure gradient was 26/13mmHg (Figure 2). Mild pulmonary valve regurgitation and moderate tricuspid valve regurgitation were recorded. Systolic pulmonary artery pressure was 45mmHg. The right heart was not dilated with FAC 53%. Left ventricle EF was 68%. MSCT showed a tumor in the pulmonary artery trunk (36x24mm) and in the left pulmonary artery 11x18mm (Figure 1). Abdominal MSCT showed normal results.

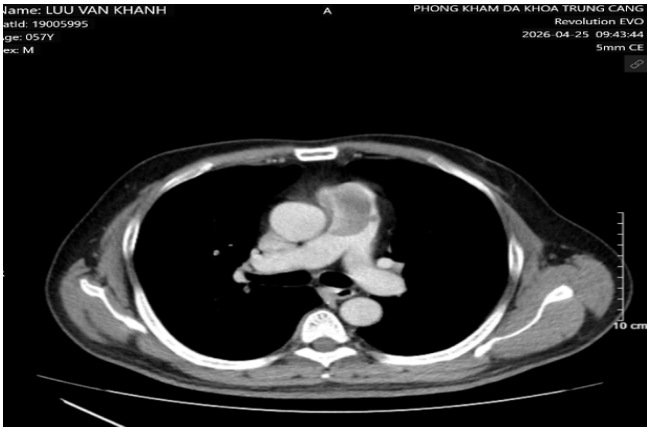

Figure 1. Tumor in the pulmonary trunk on MSCT

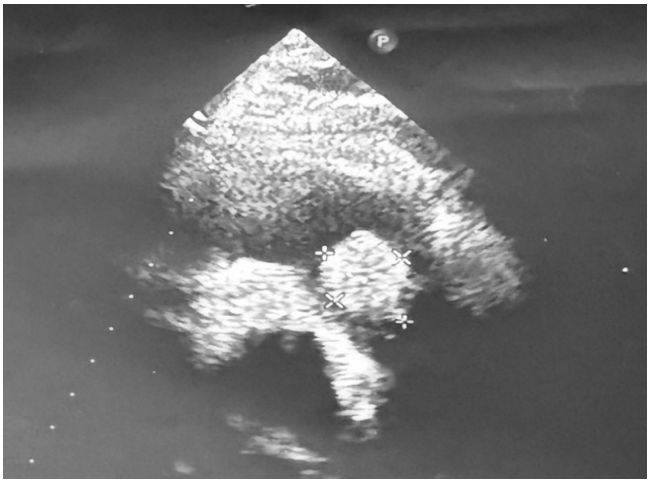

Figure 2. Tumor in the pulmonaty trunk on Echo

Surgery was performed via a median sternotomy with ascending aortic and two vena cava cannulation. After clamping the aorta and transfusing cardioplegia solution, a longitudinal incision was made in the pulmonary artery to remove the entire tumor with its main attachment from the anterior pulmonary valve leaflet (Figure 3). All three pulmonary valve leaflets were also resected and replaced with a size 25 Epic plus® biological valve. The patient was weaned off the ventilator at 10 hours and discharged after 6 days. Immuno-histochemical histopathology results showed a poorly differentiated smooth muscle sarcoma (Figure 5).

3. DICUSSION

Primary malignant tumors of the heart are rare. According to Neuville et al., intimal sarcoma is the most common type of malignant tumor of the heart (42%), followed by vascular sarcoma (26%).

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The incidence in women is almost double that in men, and more than two-thirds of cases occur in the left heart[1]. Pulmonary artery sarcoma is very rare; the literature only reports individual cases and is only diagnosed when the patient undergoes surgery [2-8]. Acounted to 1992, only 391 cases were reported worldwide with an average age of 52 ± 14 years, men accounting for 55%. The median time from the first symptom to definitive diagnosis was 100 days (47-210). 47% of cases were misdiagnosed as pulmonary embolism [4]. Most cases presented with severe symptoms such as pulmonary embolism, pericardial effusion causing compression or obstruction of the right ventricular outflow tract. Patel reported 8 cases of intimal sarcoma from the pulmonary artery, all with lung or brain metastases. Six cases underwent palliative decompression surgery, with additional chemotherapy and radiotherapy, but all died after 5-20 months, indicating a high degree of malignancy [5]. Our case was diagnosed incidentally with mild dyspnea, no pericardial effusion, no pulmonary embolism, and a relatively low pressure gradient across the pulmonary valve, in contrast to the tumor occupying almost the entire pulmonary artery trunk and left pulmonary artery during surgery.

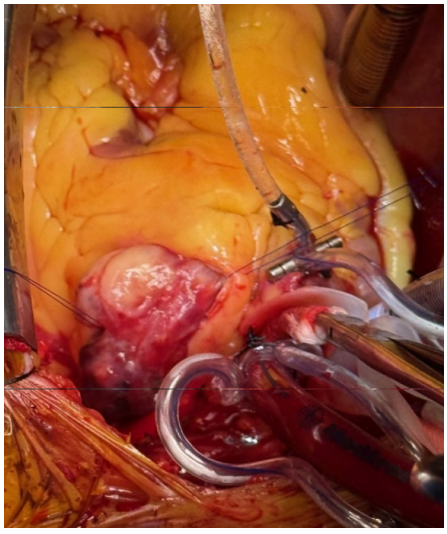

Figure 3. Tumor in the pulmonary trunk in operation

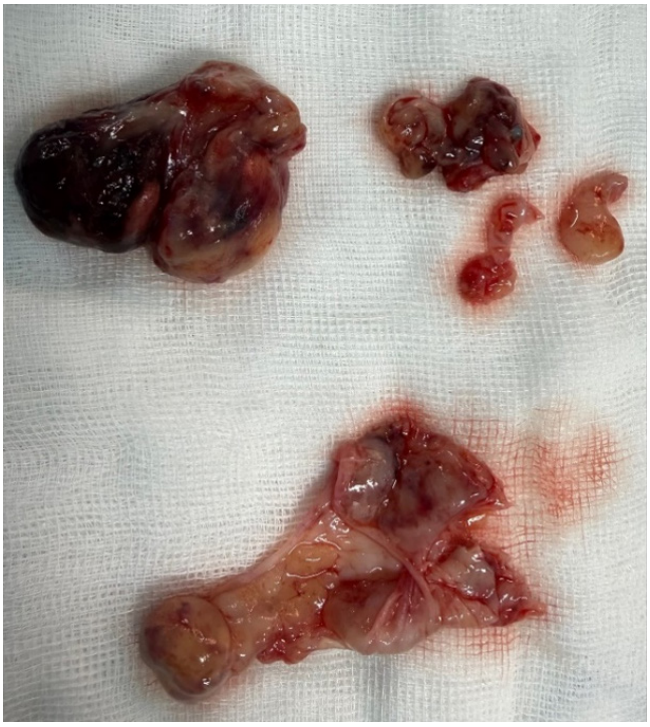

Figure 4. Tumor in pul.trunk (above) and in the left pulmonary artery (below)

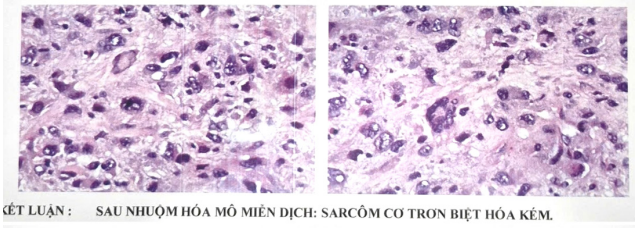

Figure 5. Results of immunohistology: poorly differentiated leiomyosarcoma

Regarding surgery, complete resection of the tumor, including all pulmonary valves and infiltrated tissues, is the preferred solution. The prognosis is quite poor, even with complete resection as mentioned above, with a 71% mortality rate in the first year and very high recurrence and metastasis rates. However, complete resection of the tumor still provides a better survival prognosis than radiotherapy or chemotherapy alone [3,5]. According to the authors, this type of tumor also responds poorly to radiotherapy and chemotherapy [3-5, 8]. Early

diagnosis is often difficult because it is not considered, and the symptoms are vague and confused with other more common diseases, especially pulmonary embolism. MSCT is a powerful tool that allows for early diagnosis and differential diagnosis of the cause.

5. CONCLUSION

Early and accurate diagnosis of malignant tumors in the pulmonary artery remains a challenge. The literature shows that complete resection of the tumor provides a better prognosis than radiotherapy and chemotherapy alone.

The authors declare no conflict of interest

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