
Photoclinic



Figure 1. Spiral CT pulmonary angiogram represents a thrombus as a filling defect in the right pulmonary artery.

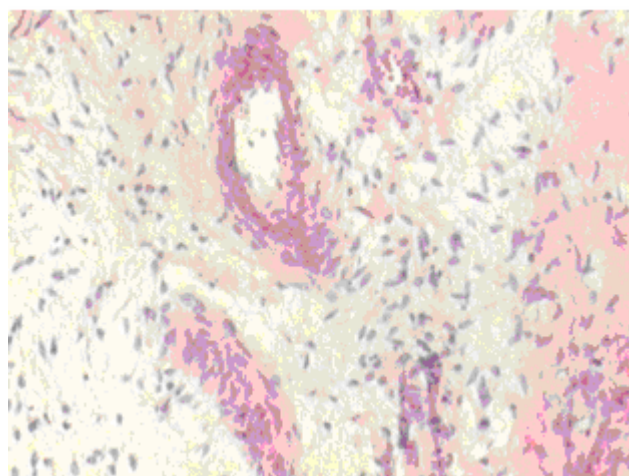


Figure 2. Histologically, polygonal spindle-shaped cells with scanty eosinophilic cytoplasm are embedded in the matrix without signs of cellular atypia or mitotic activity.

A 26-year-old man was admitted to Masih Daneshvari Hospital for investigation of brisk hemoptysis.

The disease had started around four months prior to his admission with a sudden massive bout of hemoptysis and expectoration of one-half to one glass of bloody sputum. The episodes of hemoptysis occurred every two to three days and occasionally every day. He was an otherwise healthy young man without any associated symptoms. He had no history of smoking or alcohol consumption. He was a civil engineer with no history of recent traveling. The investigations performed prior to his admission showed a normal chest X-ray, negative purified protein derivative (PPD), and several negative sputum tests for

Mycobacterium bacilla. A previous chest computed tomography (CT) had shown no pathology. Four consecutive bronchoscopic assessments revealed no pathologic findings other than a tiny blood clot situated on the left bronchial tree without any endobronchial lesions, foreign body, and/or bronchial mucosal lesions. Blood biochemistry tests, liver and kidney function tests, and blood coagulation profiles were all normal. Physical examination was unremarkable. His heart and respiratory sounds were normal. Spiral CT of the lungs with curriculum for pulmonary thrombo-embolism indicated extensive thrombosis in lobar and segmental bronchi of the right lung and segmental bronchus of the left lung (Figure 1). Subsequently, echocardiography demonstrated a large nonpedunculated mass with heterogeneous echogenicity in the right ventricle attached to its free wall. The mass was moderately mobile but could not enter the right ventricular inflow tract. Other cardiac chambers were intact.

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What is Your Diagnosis?
See the page 270 – 271 for the diagnosis

Photoclinic Diagnosis:	Right Ventricular Myxoma Presenting as Pulmonary Embolism
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The patient underwent surgery using extracorporeal circulation. Through a right ventriculotomy, the right ventricular wall was opened immediately under the valve and a yellowish 2 × 3 cm tumor appeared, which was attached to the posterior wall of the ventricle with a long pedicle. It was removed with a 2-mm margin. Then, another tumor was observed just inferior to the junction of the septal cusp and the anterior cusp of tricuspid valve, at the level of the membranous septum, which was also removed with no complication. The posterior cusp of tricuspid valve was also tumoral, which was then removed without causing any damage to the septum. Concerning the normal pressure of the pulmonary artery, no window was opened.

The operative pathology report of the three specimens labeled No: 1: “free wall”, No: 2: “A-V junction, right ventricle”, and No: 3: “tricuspid valve, posterior leaflet” was suggestive of blue and white tumoral areas with smooth glistening surface, which was histologically differentiable from organized thrombosis. Microscopically, the endocardium was replaced by ill-defined proliferation of bland-looking spindle to satellite-shaped cells in a myxoid matrix with prominent perivascular cuffing compatible with myxoma (Figure 2).

Our case shows that extensive pulmonary thromboembolism may be the only sign of a left ventricular myxoma. Similar cases have already been reported.¹⁻⁴

He had been auscultated by numerous specialists, but no auscultatory abnormality was documented. Since clinical evidence of myxoma such as heart murmur, friction rub, extrasounds, syncope, and the symptoms and signs of right heart failure were not observed in our patient, the diagnosis of myxoma was not considered.^{5, 6} Recent review articles show that half of myxomas and heart tumors, which were not detectable by auscultation or physical examination were documented by echocardiography.^{1, 4} The sensitivity of echocardiography for detection of myxoma is 100% and it seems that using such a diagnostic tool, myxoma can be detected before the abnormal auscultatory findings or obstructive symptoms are manifested.^{7, 8} At present, it is

believed that pulmonary embolism and pulmonary hypertension are common comorbidities of myxoma.^{9 - 11} Echocardiographically, the differential diagnosis of an intracardiac mass within the right ventricle includes neoplasms, thrombi, vegetations, and normal variants including embryonic remnant.¹²

The echocardiographic appearance of our case rapidly excluded all the above-mentioned differential diagnoses. The most common cardiac neoplasms are metastatic tumors, which are 20 to 40 times more common than primary tumors.¹³ The clinical manifestations of these tumors appear after documentation of the primary tumor. Therefore, in general, in a patient with cardiac neoplasm and no evidence of extracardiac tumor, myxoma is the most probable diagnosis.¹⁴⁻¹⁶

Nonetheless, in general, cardiac manifestations of extracardiac neoplasms with metastasis usually arise when background of primary cancer was recognized. Therefore, in this young man with no history of neoplastic disease, a diagnosis of primary cardiac neoplasm — the most common of which is myxoma — was immediately considered.^{7, 13}

We predicted that the intracardiac masses would be primary cardiac myxoma on a purely statistical basis. Delay in diagnosing myxoma is unfortunately common. In one study, the mean duration from onset of the symptoms to diagnosis was more than ten months and the symptoms included palpitation, fatigue, and weakness.¹⁴ Our patient presented only with hemoptysis and definite findings of pulmonary embolism, which are also rare.

At first, we believed that the patient was suffering from a type of vascular malformation; cardiac myxoma was accidentally detected by transthoracic echocardiography. The treatment of cardiac myxoma is invariably surgical, which has to be done as soon as the diagnosis is made. The patient should be followed for a long time. Magnetic resonance imaging should be performed for early detection of tumor recurrence.⁶ Ultimately, echocardiography should be undertaken early in patients with unexplained pulmonary embolism or signs of pulmonary hypertension.

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