Primary Leiomyosarcoma of the Pulmonary Artery Confirmed by Catheter Suction Biopsy*

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A patient with clinical features consistent with pulmonary embolism showed no improvement despite therapy with tissue-plasminogen activator and full-dose heparin. Transvenous catheter suction biopsy was successful in establishing an antemortem histologic diagnosis of primary pulmonary artery leiomyosarcoma. Urgent surgical intervention was performed. (CHEST 1998; 113:555-56)

Key words: catheter suction biopsy; leiomyosarcoma; pulmonary artery; pulmonary embolism

Because primary sarcoma of the pulmonary artery is extremely rare and frequently confused with other clinical conditions such as pulmonary embolism, antemortem diagnosis is seldom established.1,2 Considering the aggressive nature and critical location of the tumor, urgent surgical intervention is always mandatory and delayed diagnosis leads to a poorer prognosis.3 The first successful catheter suction biopsy of left main pulmonary artery leiomyosarcoma with partial pulmonary trunk involvement is described.

CASE REPORT

Chest pain abruptly developed in a 72-year-old woman with a 6-month history of dry cough and hemoptysis. She was diagnosed with pulmonary thromboembolism and referred to Mie University School of Medicine, First Department of Internal Medicine, by a local hospital. The patient presented with no risk factors for thromboembolism. On examination, her BP was 120/60 mm Hg; pulse, 68 beats per minute and regular; and respiration, 16 breaths per minute. A chest roentgenogram revealed left hilar enlargement and elevation of the left diaphragm, as well as infiltrates in the lower lobe of the left lung. An ECG did not detect any remarkable abnormalities. A CT scan of the chest showed only left main pulmonary arterial dilatation and small consolidations adherent to the pleura of the left lateral base. Sputum examination on three occasions failed to detect any malignant cells. Echocardiography showed neither right ventricular enlargement nor a pulmonary arterial mass. A ventilation-perfusion scan showed the absence of perfusion to the left lung with normal ventilation. Data from catheterization of the right side of the heart showed a pressure of 25/3 mm Hg for the right ventricle and a pressure of 24/8 mm Hg for the pulmonary artery. Pulmonary angiography showed an abrupt cutoff of the left main pulmonary artery, mimicking findings of pulmonary thromboembolism, but venography of the lower limbs did not reveal any deep vein thrombosis.

The patient was treated with tissue plasminogen activator and full anticoagulation. Two days later, follow-up pulmonary angiography revealed no improvement in left pulmonary arterial perfusion. Transvenous catheter suction biopsy was performed with a 10F steerable Greenfield’s embolectomy catheter (Meditech; Watertown, Mass) passed through a right femoral phlebotomy (Fig 1). There were no complications from this procedure.

Histologic examination showed bundles of spindle-shaped cells mixed with varying amounts of polygonal cells and occasional giant cells. Spindle-shaped cells with blunt-ended nuclei were arranged in fascicles or interlacing bundles (Fig 2).

![Figure 1](http://example.com/figure1.png)

**Figure 1.** Left: pulmonary angiography showed cutoff of left main pulmonary artery. Center: selective pulmonary angiography of the left lung showed a large filling defect distal to the left main pulmonary artery. Right: transvenous catheter suction biopsy was performed.
Further radiologic examinations could not reveal the source of tumor embolus and suggested that the tumor originated from the pulmonary arterial wall.

At surgery, through a median sternotomy and with full cardiopulmonary bypass, a hard tumor was found at the bifurcation of the pulmonary artery. The tumor adhered to the intima of the left main pulmonary artery and extended into the artery of the lower lobe of the left lung. Left pneumonectomy and partial resection of the pulmonary trunk was performed.

Histologic examination of the excised specimen confirmed poorly differentiated sarcoma. Immunohistochemical staining indicated leiomyosarcoma with positive reaction to smooth muscle actin using antibody against smooth muscle actin. However, reactions to cytokeratin, antimembrane antigen, factor VIII-related antigen, S-100 protein, and vimentin were all negative.

The patient remains well with no evidence of recurrence 15 months after surgery.

**DISCUSSION**

Primary pulmonary artery sarcoma is a rare and highly lethal disease. Twenty-three cases of pulmonary artery leiomyosarcoma have been reported in the world medical literature.4

Due to the critical location of the tumor mass and its malignant nature, in vitro diagnosis rarely has been reported. Although the latest developments in MRIs may differentiate between tumor and thrombus, to date there is no report on MRI appearance in primary leiomyosarcoma of the pulmonary artery. Blunn et al5 reported that MRIs can differentiate between primary leiomyosarcoma of the inferior vena cava with intraluminal growth (homogeneous intermediate signal intensity on T1-weighted images) and thrombi (hyperintense on T1- and T2-weighted images). Since the MRI appearances are variable according to the extent of organization of thrombus and coexistence of tumor and thrombus, definitive exclusion of pulmonary thromboembolism is difficult to achieve on clinical and laboratory findings. Diagnosing sarcoma of the pulmonary artery has not been considered feasible prior to surgery or autopsy.1,3,4

The poor short-term prognosis of the disease, once diagnosed, is related to delayed clinical recognition and to often inadequate surgical intervention.3,6 Survival of patients without surgical excision has been poor, regardless of adjuvant chemotherapy or irradiation. Although the case reported was suspected to be acute pulmonary thromboembolism without hemodynamic compromise, there was no absolute indication for surgical embolectomy, which would have delayed the timing of the surgical intervention if transvenous catheter suction biopsy had not been performed. Early diagnosis by this method led to a good prognosis. Talley and Franch7 have reported the only case of an intrapulmonary arterial tumor in whom catheter suction biopsy with an 8F multipurpose catheter had been successful in obtaining a histologic diagnosis. In the case reported here, a steerable Greenfield’s embolectomy catheter8 was selected to obtain a large amount of material because of its large suction cup (5 mm diameter) attached at the tip. Moreover, the steerable handle of this device enables one to achieve high accessibility to the intrapulmonary arterial mass through the chambers and the pulmonary branches of the right side of the heart. In the absence of risk factors for thromboembolic disorder and lack of response to fibrinolytic therapy, a presumptive diagnosis of malignancy should always be considered, and catheter suction biopsy is recommended to establish a definitive diagnosis of pulmonary sarcoma.

**REFERENCES**