

A Case of Pulmonary Artery Intimal Sarcoma Diagnosed with Multislice CT Scan with 3D Reconstruction

Eui-Young Choi¹, Young-Won Yoon¹, Hyuck Moon Kwon¹, Dongsoo Kim¹, Byung-Eun Park¹,
Yoo-Sun Hong², Ja-Seung Koo³, Tae-Hoon Kim⁴, and Hyun-Seung Kim¹

Division of ¹Cardiology, Department of Internal Medicine, ²Thoracic and Cardiovascular Surgery, ³Pathology, ⁴Radiology, Yonsei University College of Medicine, Seoul, Korea.

Pulmonary artery intimal sarcoma is a rare highly lethal disease, with additional retrograde extension to pulmonic valve and right ventricle being an extremely rare condition. It is frequently mistaken for pulmonary thromboembolism. We report a case of 64-year-old woman with progressive dyspnea initially suspected and treated for pulmonary thromboembolism. Her helical chest CT scan with 3 dimensional (3D) reconstruction combined with echocardiography revealed a compacting main pulmonary artery mass extending to the right ventricular outflow tract and the right pulmonary artery. After excision of the mass, the patient's condition improved dramatically, and the pathologic findings revealed pulmonary intimal sarcoma. This report emphasizes that helical chest CT with 3D reconstruction can be an important tool to differentiate the characteristics of pulmonary artery lesions, such as intimal sarcoma and thromboembolism.

Key Words: Pulmonary artery intimal sarcoma, helical CT with 3D reconstruction, echocardiography

INTRODUCTION

Pulmonary artery intimal sarcoma is a rare highly lethal disease and can be frequently mistaken for pulmonary thromboembolism. In addition, sarcoma with a retrograde extension to the pulmonic valve and the right ventricle is extremely rare. With recent technological advance, multi-slice (helical) CT scan can be performed

within just one breath-holding time and give an excellent spatial and 3 dimensional (3D) image. We report here a case that initially was suspected to be pulmonary thromboembolism, but finally pulmonary artery tumor was diagnosed applying the combination of echocardiography and helical chest CT with 3D reconstruction. Emergency operation was performed, and the findings revealed pulmonary artery intimal sarcoma originating from the main pulmonary artery (MPA), extending to the right ventricular outflow tract (RVOT) and the right pulmonary artery.

CASE REPORT

A 64-year-old woman presented symptoms of progressive dyspnea. The physical examination showed the following results: heart rate 120 bpm, blood pressure 100/60 mmHg, body temperature 37°C, respiration rate 32/min, room air oxygen saturation 90% and moderate jugular venous distension. Grade II/IV mid-systolic murmur could be heard at the left second intercostal space. A baseline ECG revealed sinus tachycardia with the right ventricular strain pattern, and the chest plain radiography showed a right ventricular enlargement with prominent pulmonary trunk. At the time of admission, there were no significant abnormal findings concerning the blood count and chemistry, except mild anemia (on the peripheral blood smear, hemoglobin 10.0 g/dL, hematocrit 32.3%) and elevated serum BNP level (827 pg/mL (reference range: 0-75 pg/mL)). D-dimer was 1.1 ug/mL (reference range: 0-0.4 ug/mL),

Received September 1, 2003
Accepted January 3, 2004

Reprint address: requests to Dr. Young-Won Yoon, Division of Cardiology, Internal Medicine, Yongdong Severance Hospital, Yonsei University College of Medicine, 146-92 Dogok-dong, Kangnam-gu, Seoul 135-720, Korea. Tel: 82-2-3497-3330/3331, Fax: 82-2-3463-3882, E-mail: drcliff@yuonc.yonsei.ac.kr

prothrombin and activated partial thromboplastin time were within the reference range. The echocardiogram performed on admission showed enlarged right ventricle (RV) with D-shaped left ventricular chamber, a compacting echogenic mass at MPA and pulmonary artery bifurcation portion (Fig. 1). Doppler test findings showed severe pulmonary hypertension, and the assumed right ventricular pressure by measuring the velocity of the tricuspid regurgitation jet was 105 mmHg. After the initial emergency evaluation, giving an impression of massive pulmonary thromboembolism, intravenous heparinization and considered intravenous thrombolytic therapy was administered. To define the precise character

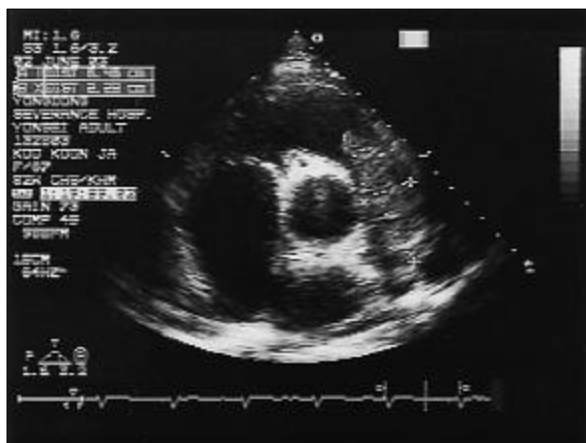


Fig. 1. Echocardiographic findings show a protruding echogenic mass impacted in the main pulmonary trunk and bifurcation portion.

of the pulmonary thrombi, helical chest CT with 3D reconstruction was done (Fig. 2). The chest CT findings demonstrated a huge intravascular mass, which contained lipid density at the MPA and RPA proximal portion, obstructing 90% of the lumen, extending to RVOT, with a minimal amount of the pericardial effusion. Due to the vague mass characterization and risk of massive distal embolization, intravenous thrombolytics could not be applied, and an emergency operation was performed. The patient was managed through median sternotomy, with total cardiopulmonary bypass and cardioplegic arrest. Operative findings revealed a hard, well capsulated whitish mass extending superiorly to the RPA and inferiorly extending to the pulmonic valve and the RVOT. The mass involving a large area of the anterior arterial wall was resected together with part of the pulmonary valve and RVOT. No significant spreading into the mediastinal fat was noted. The distal defect was reconstructed by the primary closure of the arteriotomy, and proximally, pulmonic valve anterior cusp was reconstructed with Wessex porcine pericardium (Holmbush Estate, West Sussex, England). Gross pathologic findings demonstrated a gray smooth and glistening, $11 \times 7 \times 5$ cm in size, 218 g of weight mass. On section, the surface had gelatinous gray white appearance (Fig. 3A and B). Microscopic examination of the surgical specimen revealed abundant spindle cells with hyperchromatic, pleomorphic nuclei embedded in the myxoid background,

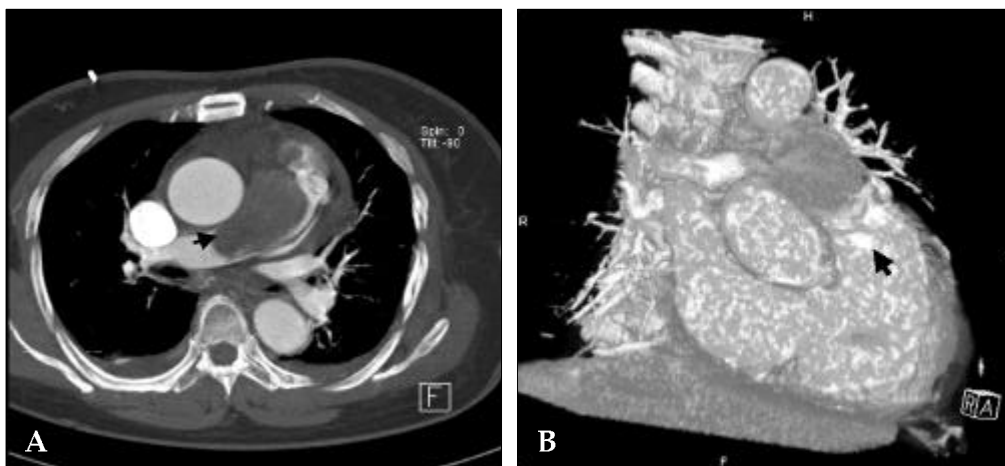


Fig. 2. Chest helical CT (A) and 3D reconstruction (B) show a right ventricular mass and saddle shaped compacting mass at the main pulmonary trunk and bifurcation portion.

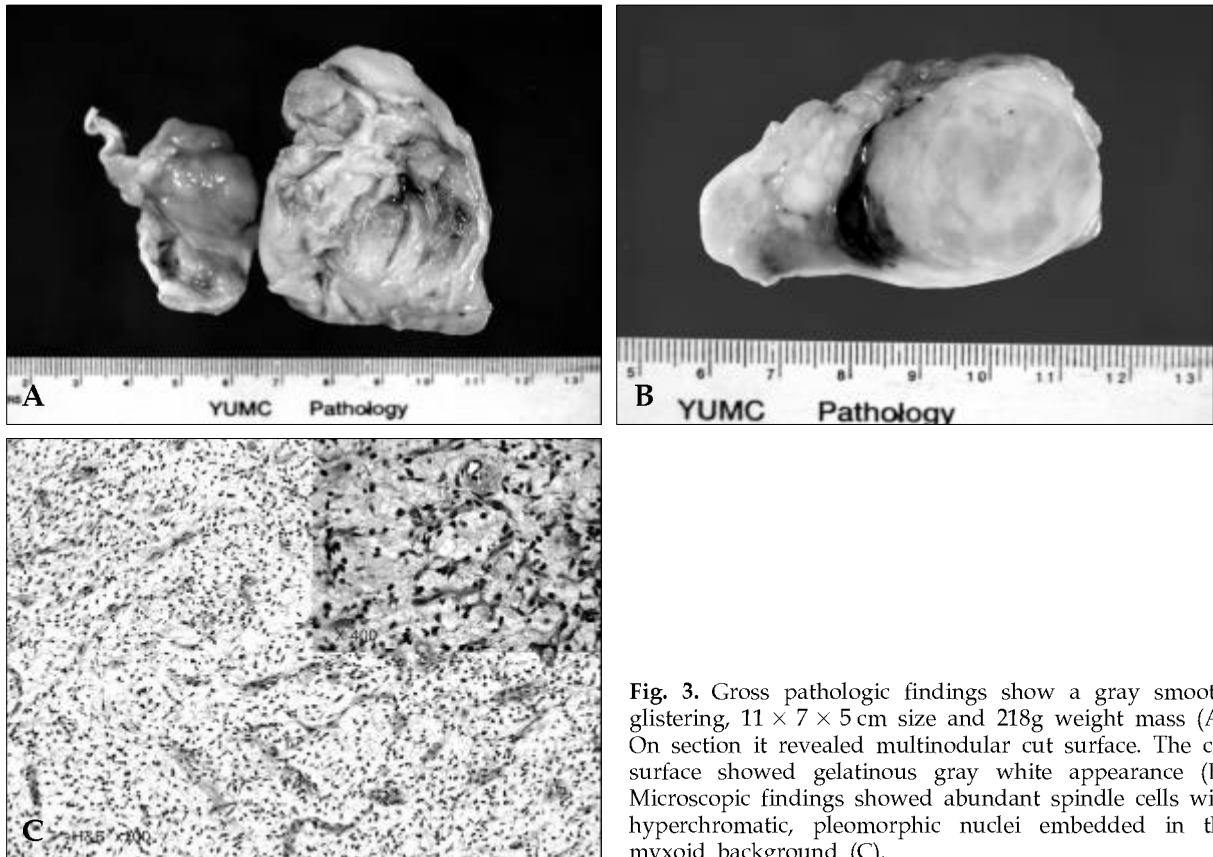


Fig. 3. Gross pathologic findings show a gray smooth, glistening, $11 \times 7 \times 5$ cm size and 218g weight mass (A). On section it revealed multinodular cut surface. The cut surface showed gelatinous gray white appearance (B). Microscopic findings showed abundant spindle cells with hyperchromatic, pleomorphic nuclei embedded in the myxoid background (C).

suggestive of intimal sarcoma (Fig. 3C). After palliative excision of the obstructing mass, the patient's condition improved dramatically. Echocardiographic findings 7 days after the operation showed no remnant mass at RVOT and pulmonary trunk; besides, the Doppler test findings showed no pulmonic regurgitation, and improvement of pulmonary artery pressure to 42 mmHg was noted.

DISCUSSION

Primary sarcomas of great vessels involve the aorta, pulmonary artery and inferior vena cava.¹ The pathologic classification of these tumors can be made on the basis of the location of the sarcoma in relation to the vessel wall, as luminal or mural.¹ Luminal sarcomas are usually intimal sarcoma, and mural sarcoma are most frequently leiomyosarcoma.¹ Primary intimal sarcoma of the pulmonary artery is a rare tumor that is most

commonly diagnosed at surgery or autopsy.² It was called as undifferentiated spindle cell sarcoma in the past.³ Important differential diagnoses include parenchymal cancer with pulmonary artery invasion and fat containing lesions, such as lipoma and hamartoma.⁴ The reported age at presentation ranges from 13 to 86 years, with the majority of cases occurring in middle age, with 2:1 female-male ratio.⁵ Pulmonary artery intimal sarcoma is thought to arise from multipotential mesenchymal cells of the intima of the vessel, and to present diverse morphologic expressions.⁵ It generally arises from the intimal layer of the right, left, and main pulmonary arteries and extends as polypoid masses into the small pulmonary arteries. As in our case, its growth in a retrograde fashion involving the pulmonary valve and right ventricle is rare. In approximately 50% patients, the tumors spread transmurally into the adjacent lung, bronchial wall or lymph nodes, and extension to myocardium and mediastinum has also been described.⁶ Systemic metastases are usually

found in the lung caused by embolic metastases but are rarely found in the liver.⁷ The prognosis is unfavorable, with the mean survival rate of 12 months after the onset of the symptoms.⁸

Because of its rarity and insidious growth characteristics, the pulmonary artery intimal sarcoma is often mistaken for pulmonary embolism, leading to inappropriate therapy, such as prolonged anticoagulation or thrombolysis.⁹ Precise preoperative diagnosis is crucial for proper exploration of the pulmonary artery, to ensure complete resection and reconstruction. Symptoms and signs such as weight loss, fever, anemia, and digital clubbing may be subtle clues to the diagnosis. Other characteristics, such as the absence of risk factors for deep vein thrombosis, high sedimentation rate, nodular parenchymal infiltrates on CT scans, unilateral absence of blood flow on perfusion scan, and lack of response to anticoagulation should give a rise to suspicion of a process other than pulmonary embolism. Unfortunately, none of the above features can exclude the possibility of chronic thromboembolic disease. Initially, our case was diagnosed as massive pulmonary thromboembolism, but after helical chest CT with 3D reconstruction, the patient was suspected to have not thromboembolism but rather pulmonary tumor.

Helical CT is a new technology leading to rapid acquisition of a large volume during the peak of intravascular enhancement. Indications are large in vascular pathology due to the possibilities of 3D reconstructions providing angiogram-like images. To assess the pulmonary artery system, this examination needs the choice of multiple parameters for acquisition (collimation, table speed, can time), injection (volume of contrast material, rate, scan delay) and reconstruction (increment, field of view). Post processing is performed on an independent workstation. Helical CT provides a reliable analysis of intracardiac and pulmonary arterial pathology.¹⁰ On CT, chronic pulmonary thromboembolic disease usually shows abrupt vascular narrowing and a cut-off instead of a continuous soft tissue filling of the pulmonary arteries as in case with the neoplasm.¹¹ Distension of the vascular lumen by the tumor and extravascular invasion into the adjacent structure can also be a clue to neoplasm, in contrast to

chronic pulmonary thromboembolism. Pathologically, pulmonary intimal sarcoma originates from subendothelial cells, which become myofibroblasts. Microscopic examination shows pleomorphic tumor with spindle and epithelioid cells, positive for actin, desmin and vimentin.¹² Literature on this disease has reported it to be uniformly fatal, with the longest survival time in one series reported to be 3.5 years, despite surgical resection. Surgical resection offers the only chance of prolonged survival. The role of chemotherapy and radiation is still uncertain. This is a contrast to pulmonary parenchymal sarcoma, which behaves less aggressively and can be cured by resection with or without adjuvant therapy.¹³

We have reported a very rare case of pulmonary intimal sarcoma originating from MPA extending to RVOT and the right pulmonary artery, which was suspected to be a pulmonary artery tumor by helical CT with 3D reconstruction and managed by palliative mass excision and pulmonary valve reconstruction. In summary, if there is persistence of the filling defect in the pulmonary artery and no response to optimal anticoagulation treatment in a patient with chronic symptoms, such as dyspnea, intimal sarcoma of the pulmonary artery should be considered in the differential diagnosis of chronic thromboembolic pulmonary hypertension.

REFERENCES

1. Brocheriou I, Quillard A, Gatecel C, Wassef M. An unusual primary vascular tumor: intimal sarcoma of the pulmonary artery. *Ann Pathol* 2000;20:69-72.
2. Pagni S, Passik CS, Riordan CH. Sarcoma of the main pulmonary artery: an unusual etiology of recurrent pulmonary emboli. *J Cardiovasc Surg (Torino)* 1999;40:457-61.
3. Johansson L, Carlen B. Sarcoma of the pulmonary artery: report of four cases with electron microscopic and immunohistochemical examinations, and review of the literature. *Virchows Arch* 1994;424:217-24.
4. Gaerte SC, Meyer CA, Winer-Muram HT, Tarver RD, Conces DJ Jr. Fat-containing lesions of the chest. *Radiographics* 2002;22:S61-78.
5. Goverder D, Pillay SV. Right pulmonary artery sarcoma. *Pathology* 2001;33:243-5.
6. Baker PB, Goodwin RA. Pulmonary artery sarcomas: a review and report of a case. *Arch Pathol Lab Med* 1985;109:35-9.

7. Shimizu J, Sasaki M, Nakamura Y, Watanabe S, Takeuchi K, Ikebata Y, et al. Simultaneous lung and liver resection for primary pulmonary leiomyosarcoma. *Respiration* 1997;64:179-81.
8. Sherrick AD, Swensen SJ, Hartman TE. Mosaic pattern of lung attenuation on CT scans: frequency among patients with pulmonary artery hypertension of different causes. *AJR* 1997;169:79-82.
9. Akomea AC, Dussek JE, Anderson DR. Pulmonary artery sarcoma mimicking pulmonary embolism: successful surgical intervention. *Ann Thorac Surg* 1996; 61:1536-8.
10. Morteale KJ, McTavish J, Ros PR. Current techniques of computed tomography. Helical CT, multidetector CT, and 3D reconstruction. *Clin Liver Dis* 2002;6:29-52.
11. Kauczor HU, Schwickert HC, Mayer E, Kersjes W, Moll R, Schweden F. Pulmonary artery sarcoma mimicking chronic thromboembolic disease: computed tomography and magnetic resonance imaging findings. *Cardiovasc Interv Radiol* 1994;17:185-9.
12. Perez MJ, Molina SR, Fresno MF, Veiga GM, Madrigal RB, Gonzalez GM, et al. An intimal sarcoma of the pulmonary artery. An immunohistochemical study. *Rev Esp Cardiol* 1998;51:850-2.
13. Mattoo A, Fedullo PF, Kapelanski D, Ilowite JS. Pulmonary artery sarcoma: a case report of surgical cure and 5-year follow-up. *Chest* 2002;122:745-7.