Spontaneously Developed Pulmonary Arterial Intramural Hematoma That Mimicked Thromboembolism

Eun-Ju Kang, MD, Ki-Nam Lee, MD, PhD, In Kim, MD, Jong-Min Chae, MD, PhD, Gun-Jik Kim, MD, PhD, Dong Heon Yang, MD, PhD, Jongmin Lee, MD, PhD

1Department of Radiology, Dong-A University College of Medicine, Busan 602-714, Korea; Departments of 2Radiology, 3Pathology, 4Thoracic Surgery and 5Cardiology, Kyungpook National University Hospital, Daegu 700-721, Korea

A 65-year-old woman visited our hospital with a complaint of acute onset dyspnea and radiological manifestations of pulmonary thromboembolism. The patient underwent an exploratory surgery to find a whitish-blue colored mass occupying almost the whole lumen of the main pulmonary arteries. Based on the pathological and radiological findings, the patient was diagnosed to have a pulmonary arterial intramural hematoma. Intramural hematomas are usually observed in the walls of the aorta, and we believe that an isolated intramural hematoma in the pulmonary artery has not been described previously.

Key words: Pulmonary artery; Intramural hematoma; CT; Ultrasound

INTRODUCTION

Intramural hematoma (IMH) is usually observed in the wall of the aorta and is characterized by the absence of a detectable intimal tear with hemorrhage to the media. Pulmonary arterial IMH is a rare event and one case was reported with an unusual complication of aortic dissection (1). To our knowledge, a report of isolated pulmonary arterial IMH has not been previously described in the literature. We report an extremely unusual case of an isolated intramural hematoma in both the pulmonary artery and the main pulmonary trunk in a 65-year-old woman with clinical manifestations mimicking a pulmonary thromboembolism.

CASE REPORT

A 65-year-old woman visited our hospital due to acute onset dyspnea. She had intermittent epigastric discomfort over the previous month and a history of healed pulmonary tuberculosis 20 years ago. Laboratory examinations showed elevated levels of CK-MB (5.0 ng/mL; normal < 3.6 ng/mL) and Troponin I (0.96 ng/mL; normal < 0.1 ng/mL). However, no abnormality was seen in the D-dimer level (180 ug/mL; normal < 326 ug/mL). The chest radiography showed a markedly decreased volume of the left lung with an architectural distortion suggestive of sequelae from old pulmonary tuberculosis, and prominent bilateral pulmonary arteries (Fig. 1A). The electrocardiogram (ECG) showed inverted T-waves at leads V1-V6, which suggested ischemia, and left axis deviation.

Transthoracic echocardiography was performed for further
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Fig. 1. 65-year-old woman visited our hospital with acute onset dyspnea.
Chest radiography (A) shows markedly decreased volume of left lung with architectural distortion and prominent bilateral pulmonary arteries. Aortic arch and descending aorta shows diffuse wall calcifications, luminal dilatation and stenosis throughout aorta. Transthoracic echocardiography (B) demonstrates significant dilatation of pulmonary trunk and huge (4.0 x 2.6 cm), solid hyperechoic mass occupying pulmonary trunk and bilateral main pulmonary arteries. Contrast-enhanced chest CT (C) documents presence of homogeneous (about 75 HU of density) mass with near total luminal obstruction of bilateral pulmonary arteries (not shown in this figure). Cardiac magnetic resonance imaging (MRI) reveals that mass is heterogeneously iso to high signal intensity on both T1- (D) and T2-weighted images (E). Also noted, capsule-like high signal intensity (arrows) at boundary with pulmonary arterial lumen.

evaluation of cardiac function. This revealed a diffusely distended pulmonary trunk and a huge (4.0 x 2.6 cm), capsulated homogeneously hyperechoic mass within the pulmonary trunk and the bilateral main pulmonary arteries (Fig. 1B). The pulmonary arterial pressure was measured at 67 mmHg, which is indicative of severe pulmonary hypertension. In addition, a dilated ascending aorta, right atrium and right ventricle were observed. The impression of the echocardiography was reported as pulmonary arterial thromboembolism.

Subsequently, contrast-enhanced chest computed tomography (CT) was performed to confirm the pulmonary embolism, and the mass in the pulmonary artery showed homogeneous opacity with 75 HU on the post-contrast enhancement scan (Fig. 1C). The mass extended into the bilateral main pulmonary arteries with near-total luminal obliteration. There was no evidence of obstructive lesion in the veins of both lower extremities. Cardiac magnetic resonance imaging (MRI) displayed the mass as having heterogeneously iso- to hyper-intense signals on the T1 and T2-weighted images with a remarkable capsule-like hyperintense margin facing the pulmonary arterial lumen (Fig. 1D, E). The mass showed a well-defined mural margin without evidence of extravascular extension beyond the pulmonary arterial wall. The diagnosis by CT and MRI was a non-malignant pulmonary artery tumor without exclusion of chronic thromboembolism.

Since her dyspnea was rapidly aggravated in spite of highly concentrated oxygen inhalation, an exploratory surgical operation was conducted by thoracic surgeons. During the operation, a whitish-blue colored mass was exposed by incision on the cranial aspect of the pulmonary trunk bifurcation. The mass occupied almost the whole lumen of the bilateral main pulmonary arteries and it was
encapsulated by a membranous structure (Fig. 1F). No defect was observed on the capsular structure, which was continuous to the adjacent intima. The surgeons’ initial on-site impression was a neoplasm. During a trial of excision, a mixed stage blood clot (red and white color clots) effused from the mass. A piece of the free wall of the mass was collected and sent for pathological exam; as many of the contents as possible were removed from the mass, either by aspiration or picking, and the defect of the free wall was sutured. Histological evaluation of the capsular tissue revealed positive special staining for both smooth muscle antigen (SMA) and elastic fibers, which was suggestive of arterial wall (Fig. 1G). Based on the pathological and radiological findings, the patient was diagnosed as having pulmonary arterial IMH with pulmonary hypertension.

**DISCUSSION**

Most of the occupying lesions in the pulmonary arteries are pulmonary thromboembolisms or intimal sarcomas and the differential diagnosis is usually made by CT number or signal intensity of each of the MR sequences. Pulmonary arterial IMH is a rare event and it is an unusual complication of aortic dissection (1). To the best of our knowledge, isolated pulmonary arterial IMH has not been previously reported. Also, the preoperative diagnosis of the pulmonary IMH was very difficult, because the radiologic features of the lesion in the CT and MRI were very similar to a pulmonary thromboembolism.

We presumed the cause of the pulmonary IMH is the same as that of pulmonary arterial dissection. According to the previously reported cases of pulmonary arterial dissection, most commonly, the patients had an underlying cardiac disease such as a patent ductus arteriosus or rheumatic mitral stenosis (2, 3). Pulmonary arterial dissection associated with idiopathic pulmonary hypertension or a connective tissue disorder has been reported (4).

Our patient had severe parenchymal destruction of the left lung as sequelae of old pulmonary tuberculosis, which may have caused pulmonary hypertension, similar to the report by Lee et al. (5). In addition, the patient’s aorta showed diffuse massive transmural calcifications as well as luminal dilatation with stenosis throughout the aorta (Fig. 1A, C). These findings suggested underlying chronic autoimmune arteritis, such as Takayasu’s arteritis, which may have had an influence on the architectural integrity of the pulmonary arterial wall. Since the postoperative mentality of the our patient decreased to a semicoma state and no past medical records existed in our institute, additional information to confirm Takayasu arteritis (6) could not be acquired.

We report on an extremely unusual case of an isolated IMH in the pulmonary artery. We propose that in the case of the mass-like lesions located in the pulmonary trunk, if the mass shows the same density or signal intensity as hematoma and capsule-like margin, then pulmonary IMH should be included in the differential diagnosis.
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