Serial CT Findings of Pulmonary Artery Intimal Sarcoma in 4 Months: a case report

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Intimal sarcoma of pulmonary artery is a disease easily misdiagnosed as acute or chronic pulmonary thromboembolism. Although the disease is rarely seen, life-saving surgical treatment can be applied if correctly diagnosed. We report a case of 69-year-old woman with clinical presentation of progressive dyspnea received medical treatment because pulmonary embolism was initially diagnosed. But CT study 4 months later showed progression and contrast enhancement of the supposed pulmonary “thromboembolism”. Endarterectomy was performed and pathology showed intimal sarcoma. The patient expired 4 days after operation due to hepatic failure which was considered to be resulted from chronic pulmonary hypertension exacerbating due to circulatory arrest for 3 times with intermittent reperfusion during the surgery. This report provides the diagnostic value of CT study with the enhancement of tumor and expansile appearance of interlobar arteries.

Since Mandelstamm’s first description in 1923, there have been more than 200 cases of pulmonary arterial sarcoma reported in the English literature. Most tumors appear as filling defects in the pulmonary artery or its tributaries on enhanced computed tomography (CT) scans [1]. Because of its rarity pulmonary arterial sarcoma is often misdiagnosed as chronic pulmonary thromboembolism and results in delayed surgical treatment, lower survival rate and poor prognosis. Computed tomography is useful to detect pulmonary arterial intimal sarcoma. We report a case with intimal sarcoma of pulmonary artery and its serial CT findings.

CASE REPORT

A 69 year-old woman suffered from chest pain for four days. Chest plain film (Fig. 1) showed prominent right pulmonary trunk and right lower lung consolidation. There is right lower lung atelectasis, consolidation and right pleural effusion. Chest CT (Fig. 2a) showed intraluminal filling defect in right pulmonary artery and pulmonary artery of right lower lobe. Bilateral lower limb color Doppler sonographic survey for deep vein thrombosis revealed negative finding. Warfarin was then used to control pulmonary embolism. Four months later, chest radiography (Fig. 2b) showed the right hilum in a “cloverleaf” appearance. Follow up chest CT (Fig. 2c) showed aggravated condition of the filling defect with both antegrade and retrograde extension and contrast enhancement arising from the arterial wall. There were also prominent expansile right pulmonary artery and its branches in lung parenchyma. No pulmonary emboli were noted in left pulmonary arteries. Endarterectomy of right pulmonary artery was performed. Tumor in pulmonary arteries of right upper lobe and right middle lobe was removed. Microscopically, there were polygonal epithelioid tumor cells containing enlarged round to oval vesic-
ular nuclei with prominent nucleoli, atypical mitoses and areas of tumor necrosis. Immunopositivity for vimentin was evident (Fig. 3). Intimal sarcoma of pulmonary artery was diagnosed. Unfortunately, elevated blood bilirubin level and liver enzymes were noted since the first post-operative day. The patient died four days post operation due to hepatic insufficiency. The cause of death was considered to be resulted from chronic pulmonary hypertension and hepatic venous congestion which exacerbated due to circulatory arrests for 3 times with intermittent reperfusion during the surgery.

**DISCUSSION**

Pulmonary arterial intimal sarcoma is thought to arise from the intimal layer of the right, left and main pulmonary arteries and extends as polypoid masses into the small pulmonary arteries [2]. Because of its rarity and insidious growth characteristics, the pulmonary artery sarcoma is often misdiagnosed as pulmonary embolism, leading to inappropriate therapy, such as prolonged anticoagulation or thrombolysis [3]. Other characteristics, such as the absence of risk factor for deep vein thrombosis, high sedimentation rate, unilateral absence of blood flow on perfusion scan, and lack of response to anticoagu-

**Figure 1.** Chest plain film showed consolidation in the right lower lung (arrowhead), blunting of right costophrenic angle. Right pulmonary artery (short arrow) is more prominent than the left side (long arrow).

**Figure 2.** a. Initial CT examination: A non-enhanced filling defect is noted in distal right pulmonary artery (long arrow) and proximal right lower lobe artery (short arrow), characteristic appearance of thromboemboli. b. Chest radiography showed consolidation of the right hilum (arrow) with a “cloverleaf” appearance. c. Follow up chest CT images 4 months later: Enhancement (arrowhead) can be observed within the filling defect in right pulmonary artery (long arrow), right lower lobe and segmental arteries (short arrow), abutting the arterial wall. Expansile arteries and resolved right pleural effusion are also evident.
Intimal sarcoma of pulmonary artery

lation should give rise to suspicion of a process other than pulmonary embolism [4]. An important but rare finding was the expansion of any portion of the involved pulmonary artery. This finding was seen in only one of forty patients (3%) with embolic disease at the level of the interlobar artery [1].

As in our case, CT showed enhancement within the supposed thrombus and the filling defect arising from the arterial wall with antegrade and retrograde extension. Expansile central pulmonary artery and interlobar arteries are also evident. These CT features are all suggestive of pulmonary artery sarcoma [5].

Gadolinium-enhanced MR imaging of the pulmonary arteries had been used to differentiate blood clot from tumor [6]. But enhanced lesion arising from unilateral arterial wall and expansile central pulmonary arteries in CT study are strongly suggested to indicate the diagnosis of intimal sarcoma rather than thromboembolism [7]. The mosaic pattern on high-resolution CT caused by pulmonary arterial intimal sarcoma was reported [8] and indistinguishable from chronic thromboembolic pulmonary artery hypertension.

In conclusion, pulmonary arterial intimal sarcoma is uncommon and may be mistaken for pulmonary emboli. The possibility of this diagnosis should be considered when evaluating an enhanced thrombus with antegrade and retrograde extension and expansile arterial diameter without contralateral abnormalities. Correct diagnosis and early surgical intervention are considered to be the best life-saving methods.

**REFERENCES**

2. Goverder D, Pillay SV. Right pulmonary artery sarcoma. Pathology 2001; 33: 243-245

**Figure 3.** Microscopic study showed a pleomorphic tumor with spindle and epithelioid cells (long arrow). Note immunopositivity for vimentin (short arrow) both for tumor cells and endothelial cells.
肺動脈內膜肉瘤之連續四個月電腦斷層變化：
病例報告

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肺動脈內膜肉瘤是一個容易被誤診為急慢性肺動脈栓塞的疾病。此疾病雖然罕見，但是如果可以正確診斷的話就可以及早施行手術治療。我們報告一位 69 歲女性病患起初表現出漸進性的呼吸困難並被診斷為肺動脈栓塞，經四個月的抗凝血剤治療後，胸部電腦斷層顯示出更大的肺動脈血栓且施打顯影劑後有部分顯影的情形，經手術取出血栓後病理科證實為肺動脈內膜肉瘤。病患於開刀 4 天後死於肝衰竭。我們在此介紹此疾病在四個月內的連續電腦斷層變化。