

Case Report

Idiopathic intimal sarcoma of the pulmonary artery: report of a case

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Received April 18, 2017; Accepted April 26, 2017; Epub August 1, 2017; Published August 15, 2017

Abstract: We report on a 51 years old woman patients with pulmonary artery intimal sarcoma, whose first symptoms was paroxysmal cough without obvious cause, and with left chest pain. It is misdiagnosed as Pleural effusion, cryptogenic organizing pneumonia and pulmonary artery embolism with pulmonary infarction successively. After pulmonary artery endarterectomy and tumor biopsy, histopathology revealed right pulmonary artery intimal sarcoma. And then she was treated by radiotherapy and chemotherapy, and died 32 months after operation.

Keywords: Pulmonary artery intimal sarcoma, misdiagnose, radiotherapy and chemotherapy

Introduction

The incidence rate of intimal sarcoma of the pulmonary artery is very rare, firstly described at autopsy by Mandelstamm in 1923 [1], and since then, fewer than 250 cases have been reported [2]. It is usually misdiagnosed as pulmonary artery thrombosis because of no specific clinical symptoms. The survival rate of pulmonary artery intimal sarcoma is normally low; the mean survival time was usually less than 2 months for those without surgical resection, and approximately 10 months for that surgical treatment [3]. Compared with chemotherapy and radiotherapy, operation therapy can alleviate symptoms and lengthens the patient's life. Here we report a case of 51 years old woman patients with pulmonary artery intimal sarcoma.

Case report

A 51-year-old woman presented with a chief complaint of paroxysmal cough and with left chest pain. Computed tomography (CT) of the chest showed right pneumonia with less pleural effusion. The above symptoms were relieved after Intravenous drip by penicillin, cefazolin and levofloxacin lactate. 3 month later, serious cough and chest pain appeared. It is diagnosed

as Pleural effusion, cryptogenic organizing pneumonia after group consultation by several hospitals. Physical examination revealed tachypnea (20 breaths per minute), a pulse of 84 beats per minute. Blood pressure was 100/80 mm Hg, and body temperature was 36.2°C. Enhanced CT showed the right bottom pulmonary artery thrombus (**Figure 1**), then diagnosed as pulmonary artery embolism with pulmonary infarction. But the illness was not become well by one year oral warfarin anticoagulant therapy.

The pulmonary artery endarterectomy and tumor biopsy were performed, displaying fish like tissue in the vascular cavity with no limits of vascular wall. The microscope showed irregular arranged spindle cells with pleomorphic nuclei in a myxoid and collagenized background (**Figure 2**). The immunohistochemistry showed that positive staining were smooth muscle actin, CD34, vimentin and desmin protein (**Figure 3**), but the negative staining were MyoD-1, VEGF, EGFR, TTF-1, laminin, EMA, CD10 and CD31. On the basis of morphological and immunohistochemical findings, it is finally diagnosed as right pulmonary artery intimal sarcoma. The patient died 32 months after surgery and radiotherapy because of respiratory failure and heart failure due to tumor progression.

Case rare idiopathic intimal sarcoma of the pulmonary artery

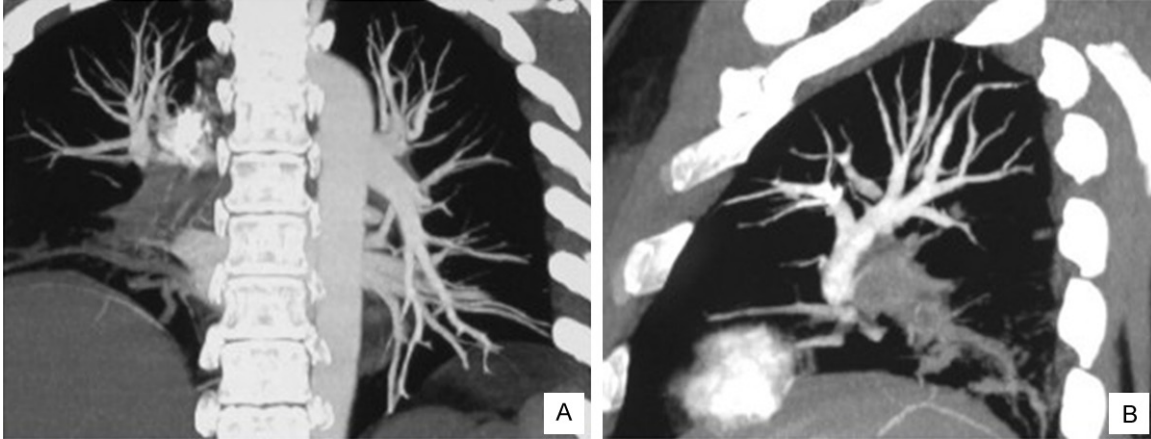


Figure 1. CT shows the right bottom pulmonary artery thrombus. A: Coronal; B: Sagittal.

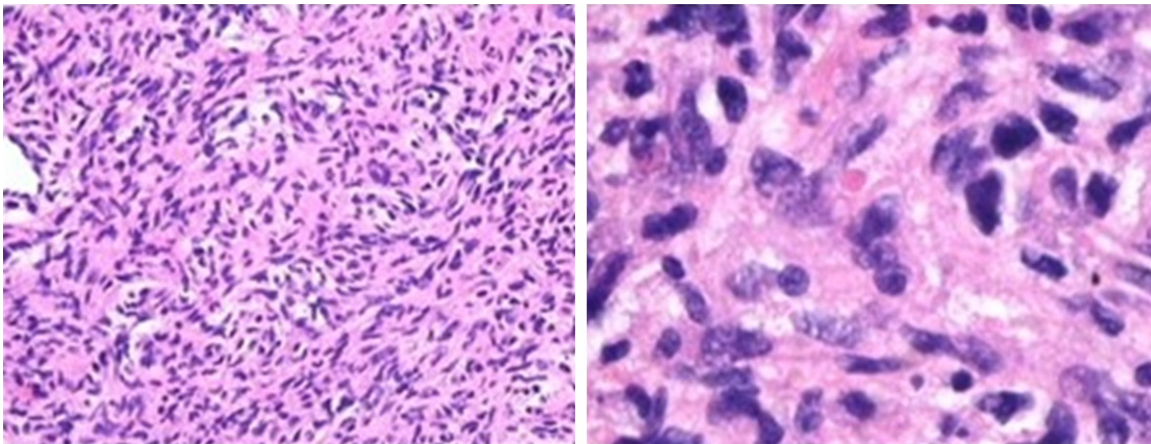


Figure 2. The microscope showed Irregular arranged spindle cells with pleomorphic nuclei in a myxoid and collagenized background (H&E stain, $\times 100$ and $\times 400$).

Discussion

Intimal sarcoma is a rare tumor that arises from large vessels, including the aorta and pulmonary artery. Pulmonary artery intimal sarcoma is lack of specific clinical manifestations [4, 5]. The most patients have chest pain, cough, hemoptysis, shortness of breath, syncope, fever and fatigue. On base of the lack of specific symptoms, it is easily to be misdiagnosed [6]. In our report, the woman presented with a chief complaint of paroxysmal cough and with left chest pain, and firstly diagnosed as pneumonia and pulmonary artery embolism, resulting in delayed diagnosis and treatment.

Pathological examination could offer confirmed evidence for the diagnosis of pulmonary artery intimal sarcoma [7]. Histopathology revealed

irregular arranged spindle cells with pleomorphic nuclei in a myxoid and collagenized background [8]. The immunohistochemistry showed that positive staining were smooth muscle actin, CD34, vimentin and desmin protein. It is pointed out that these cancer cells have the ability of differentiation into leiomyosarcoma, angiosarcoma and rhabdomyosarcoma.

The survival time of pulmonary artery intimal sarcoma is usually short, an average of 13 to 18 months. Operation is the first choice of treatment. In this report, the patient was treated by radiotherapy and chemotherapy, and died 32 months after operation. So the surgical resection, combined with radiotherapy and chemotherapy, if possible, is known to be the best diagnosis and treatment option for patients with pulmonary artery sarcoma.

Case rare idiopathic intimal sarcoma of the pulmonary artery

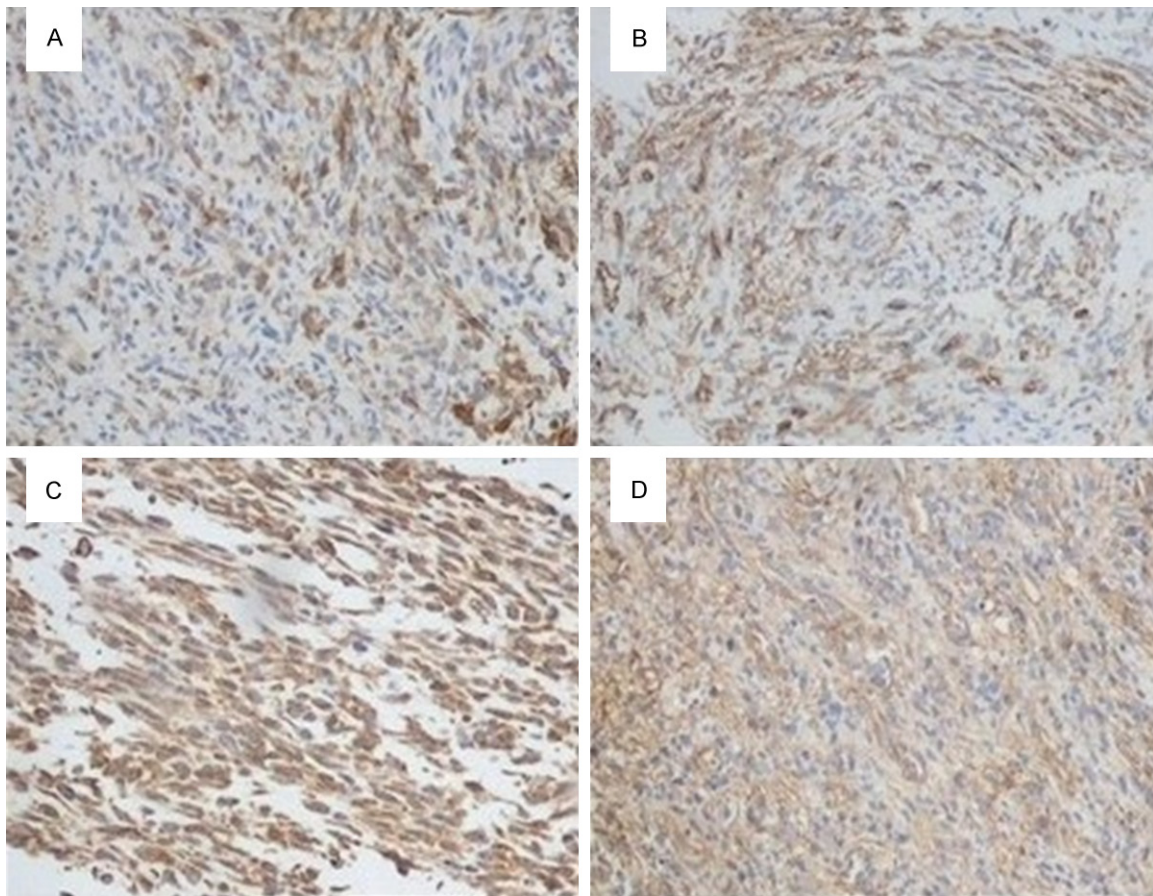


Figure 3. The immunohistochemistry showed that positive staining were (A) smooth muscle actin; (B) CD34; (C) vimentin and (D) desmin protein.

Acknowledgements

The authors declare that they have no competing interests. This project was funded by the Key Disciplines at the Zhejiang Provincial and Shaoxing city level (#GJSX-010-003).

Disclosure of conflict of interest

None.

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References

- [1] Mandelstamm M. Uber primary neubildungen des herzen. *Virchows Arch Pathol Anat* 1923; 245: 43-54.
- [2] Choi YM, Jang EK, Ahn SH, Jeon MJ, Han JM, Kim SC, Han DJ, Gong G, Kim TY, Shong YK, Kim WB. Long-term survival of a patient with pulmonary artery intimal sarcoma after sequential metastasectomies of the thyroid and

adrenal glands. *Endocrinol Metab (Seoul)* 2013; 28: 46-49.

- [3] Funatsu Y, Hirayama M, Shiraishi J, Asakura T, Wakaki M, Yamada E, Fujimoto K, Satomi R, Inaki S, Murata Y, Oyamada Y. Intimal sarcoma of the pulmonary artery treated with pazopanib. *Intern Med* 2016; 55: 2197-202.
- [4] Umezawa K, Takamura K, Yamamoto M, Kikuchi K. Pulmonary artery intimal sarcoma. *Am J Respir Crit Care Med* 2014; 190: 67-68.
- [5] Hu W, Xie Y, Zhang DD. Pulmonary artery intimal sarcoma diagnosed by percutaneous transcatheter aspiration. *Chin Med J (Engl)* 2013; 26: 1590-1591.
- [6] Sakata H, Suzuki K, Furuse H, Taniguchi H. Pulmonary artery intimal sarcoma invading the left lung. *Intern Med* 2016; 55: 1397-1398.
- [7] Evison M, Crosbie P, Chaturvedi A, Shah R, Booton R. Pulmonary artery sarcoma: a rare thoracic tumor frequently misdiagnosed at presentation. *Thorac Cancer* 2015; 6: 797-799.
- [8] Attinà D, Niro F, Tchouanté P, Mineo G, Russo V, Palazzini M, Galiè N, Fanti S, Lovato L, Zompatori M. Pulmonary artery intimal sarcoma. Problems in the differential diagnosis. *Radiol Med* 2013; 118: 1259-1268.