Recurrent intimal sarcoma mimicking pulmonary embolism

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A 58-year-old man presented with increasing cough, chest pain and shortness of breath over a period of 4 weeks. Chest computed tomography (CT) identified a mass in the right hilum obstructing the right pulmonary artery without pleural effusion (Fig. 1A). The definitive diagnosis of pulmonary artery intimal sarcoma was made using needle biopsy.
In March 2009, he received combination chemotherapy comprising ifosfamide plus epirubicin and radiotherapy with a total dose of 66 Gy. Forty-two months after initial treatment, there was evidence of recurrence as a pulmonary metastasis in the left upper lobe (Fig. 1B). The recurrence was successfully treated with radiotherapy. In March 2014, a right pulmonary mass was treated with radiotherapy. After the last course of radiotherapy, he had a 9-month disease-free interval with no signs of recurrence of the intimal sarcoma.

In December 2014, chest CT was performed as a routine evaluation to identify any metastatic lesions. Although the patient had no symptoms or complaints, contrast-enhanced CT of the chest revealed a homogeneous filling defect of the main pulmonary artery into the proximal left branch (Fig. 1C). $^{18}$F-fluorodeoxyglucose (FDG) positron emission tomography revealed a weak accumulation within this lesion, showing a maximum standardized uptake value ($SUV_{\text{max}}$) of 3.3 (Fig. 1D). Although not particularly high, this $SUV_{\text{max}}$ was thought to be significantly higher than the typical value obtained from $^{18}$F-FDG accumulation in a thrombus, suggesting the lesion was a metastasis.

The patient underwent surgical resection of a fleshy tumor within the main pulmonary artery. The histological diagnosis was recurrent intimal sarcoma, not pulmonary embolism (Fig. 1E). Histologically, the tumor within the pulmonary artery was identified as a metastasis arising from the original intimal sarcoma, because the histological morphology and immunohistochemical pattern were similar to those of the original intimal sarcoma. The patient was discharged 10 days after surgery.

Pulmonary artery intimal sarcoma is extremely rare, and is often misdiagnosed as pulmonary embolism. In our case, ~4 years after the initial diagnosis of intimal sarcoma, a recurrence of intimal sarcoma was identified as a tumor within the pulmonary artery. It may be difficult to discriminate between pulmonary embolism and a recurrent intimal sarcoma; however, we should suspect a potential recurrence of intimal sarcoma when the patient has had a prior diagnosis of intimal sarcoma.