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Low-grade sarcoma of the right upper lobe vein mimicking a metastatic disease

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We present a case of a 23-year-old patient with a primary tumor of the right upper vein and endocavitary expansion in the left atrium associated with concomitant infiltrative-like lesions of the upper and middle lobes. These lesions were suspected to be metastatic. The richly vascularized tumor was completely removed on cardiopulmonary bypass through a right thoracotomy. The pathologic findings showed a low-grade sarcoma invading the left atrium. The clinically suspected pulmonary metastases were in reality venous infarcts of the right upper and middle lobes. A complete obstruction of the pulmonary veins may create intrapulmonary lesions that can mimic metastatic lesions and should not exclude surgical treatment.

CLINICAL SUMMARY

A 23-year-old man presented with recurrent hemoptysis after a pulmonary infection. The chest radiograph showed opacity of the right upper lobe. Chest computed tomography (CT) demonstrated a sizeable left atrial mass originating from the right upper vein (Figure 1, A) and 2 lesions in the right upper and middle lobes (Figure 1, B). Tumor markers were negative. Transesophageal echography showed a 5 × 4–cm tumor occluding the right upper vein without obstruction of the mitral valve. The tumor did not completely invade the wall of the left atrium. F-18 fluorodeoxyglucose positron emission (PET) CT showed an intense fixation of the tumor with a moderate fixation of the pulmonary lesions (Figure 1, C). Maximum standardized uptake values were 4.39 for the left atrial tumor and 2.68 and 2.07 for pulmonary lesions. The CT angiography showed total occlusion of the right upper pulmonary vein (Figure 1, D). Preoperative heart catheterization showed a normal cardiac index and normal pulmonary pressure. Capillary pulmonary pressure showed a mean gradient of 6 mm Hg between the left and the right pulmonary sides.

The operation was performed through a right posterolateral thoracotomy in the fifth interspace. After extrapleural dissection required by hypervascularized adhesions, a consolidated and retracted upper lobe, an enormous upper lobe vein, and the absence of extravascular tumor were observed. Normothermic cardiopulmonary bypass was established between both venae cavae and ascending aorta. Under aortic clamp and myocardial protection, the left atrium was opened around the venoatrial junction allowing en bloc removal of the upper bilobe and the intra-atrial tumor, which was floating free in the cavity. Reconstruction of the left atrium consisted of a simple plasty without narrowing the lower lobe vein. The bypass was stopped after deairing the left cavities, and the left atrium margins were tumor-free on frozen section (Figure 2, A).

The patient was extubated at the end of the procedure, had an uneventful postoperative course, and was discharged on day 14. Pathologic examination of the tumor identified a low-grade myxoid sarcoma (Figure 2, B). The mitotic index activity measured by Ki-67 immunochemistry dosage was 10%.

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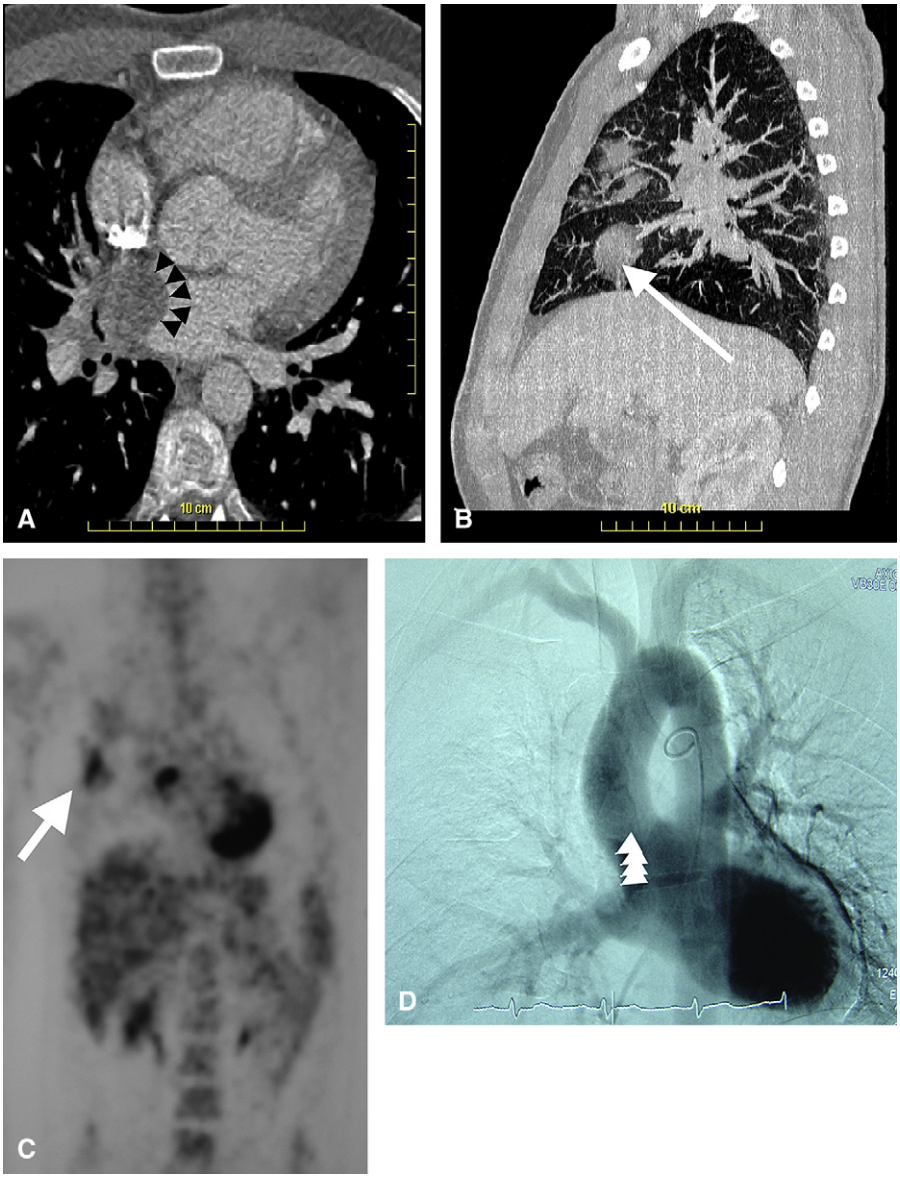

FIGURE 1. A, Preoperative chest computed tomography (CT) scan showing a solid mass in the left atrium with an occlusion of the right upper pulmonary vein *(small black arrows)*. B, Preoperative chest CT demonstrating 2 masses in the upper *(black arrow)* and middle *(white arrow)* lobes. C, F-18 fluorodeoxyglucose positron emission CT showing an intense fixation of the tumor with a moderate fixation of the pulmonary opacities. D, Angiography showing complete occlusion of the right upper vein *(white arrows)*.

DISCUSSION

Primary malignant tumors of the pulmonary vein are rarely described.[1,2] These tumors expand usually to the left atrium. A tumor of the left atrium is potentially lethal because of intracavitary or mitral obstruction, peripheral embolization, and rhythm disturbances. Therefore, surgery should be performed as soon as possible after a cardiac tumor is identified.[3]

In the present case, surgery was discussed because the pulmonary-associated lesions were suspected to be metastatic. Park and colleagues[4] consider that surgery should be proposed for metastatic patients when cardiovascular collapse is thought to be imminent or when few treatment alternatives exist, as in this case of such a young patient.[4] Despite the radiologic aspect of the pulmonary nodules on chest CT and their fixation on the PET CT scan, which resembled possible metastatic lesions, final histology demonstrated a venous pulmonary infarction, which may occur from a sudden occlusion of a pulmonary vein branch. However, the resection required a bilobectomy or a pneumonectomy because of the hilar location of the primary tumor. The surgical challenge was to preserve the lower lobe for this young patient.

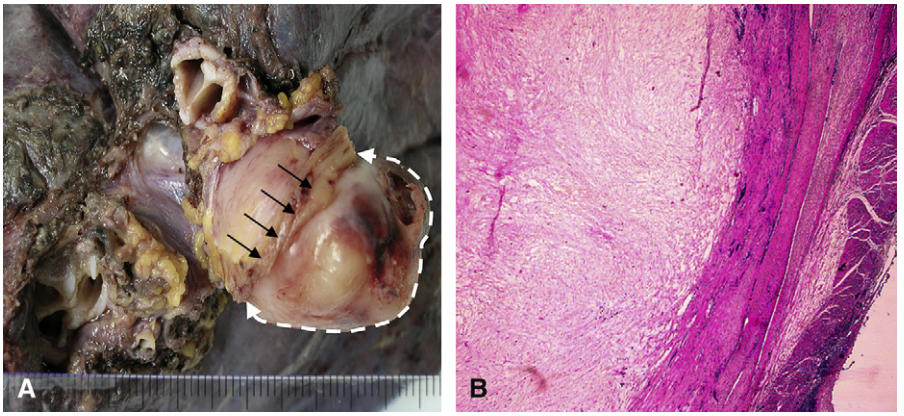

FIGURE 2. A, Operative view of the upper bilobectomy extended to the left atrium *(black arrows)* and the intracavitary malignant tumor invading the pulmonary vein *(white arrow)*. B, Histologic aspect of the tumor and the left atrial wall (intermediate magnification with hematein and eosin stain).

Tumors of the heart are extremely uncommon and almost all are benign, namely myxomas located in the left atrium. Almost all primary malignant tumors are sarcomas. The level of mitotic activity is the most important predictor of survival.[5] This tumor was a primary, low-grade sarcoma of the pulmonary vein. The low level of mitotic activity in this patient seemed a positive prognosticator. In this setting, an en bloc resection represented the gold standard for the observed pathologic extension of the disease.[1]

In the presence of an occluded pulmonary vein and concomitant suspicious pulmonary lesions, the possibility of multiple venous infarctions should be considered, thereby reinforcing the indication for a surgical option.

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