Pulmonary Artery Sarcoma Diagnosed by Endobronchial Ultrasound-Guided Transbronchial Needle Aspiration

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Pulmonary artery sarcoma (PAS) is a rare tumor that is often detected at an advanced stage, when disease is so widespread that a radical surgical procedure is no longer indicated. Therefore, less invasive biopsy techniques are required to establish a definitive preoperative diagnosis. Endobronchial ultrasound (EBUS) is useful for producing real-time images of both lymph nodes and the interior of pulmonary arteries adjacent to the bronchi. We report a case with masslike lesions in the pulmonary artery that were observed by EBUS and from which tissue was obtained by endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) to establish a diagnosis of PAS.


Previously, most cases of pulmonary artery sarcoma (PAS) were diagnosed at autopsy or during operation. However, in the past decade, new imaging techniques have increased the rate of preoperative detection of this disease. Given that the tumor is so widespread in many patients that a radical surgical procedure is no longer indicated, less invasive biopsy techniques are required to establish a definitive preoperative diagnosis.

Recently, a new bronchoscopic ultrasound technique has emerged: convex probe endobronchial ultrasound (CP-EBUS) uses a device with a curvilinear electronic transducer on the tip of a flexible bronchovideoscope, which allows real-time EBUS-transbronchial needle aspiration (TBNA) [1]. Although the main indication for EBUS-TBNA is lymph node staging, it can also be used to diagnose intrapulmonary tumors, unknown hilar lymphadenopathy or mediastinal lymphadenopathy or both, and mediastinal tumors. In the present case, EBUS-TBNA was used to diagnose PAS.

A 63-year-old man presented with dry cough that had lasted 2 weeks. Chest roentgenogram showed a nodular lesion in the right upper lobe of the lung. Enhanced computed tomography (CT) of the chest confirmed this nodular lesion and multiple mass lesions in the vascular lumen of the bilateral pulmonary arteries (PAs) that spread from the right ventricular outflow tract to the bilateral main PAs and the left lower lobe tract of the PA (Fig 1). To differentiate between pulmonary embolism and malignant disease, positron emission tomography (PET)-CT was performed. Fluorodeoxyglucose (FDG) uptake was observed in the multiple mass lesions in the bilateral pulmonary arteries and the nodular lesion in the right upper lobe of the lung (Fig 2). The maximum standardized uptake value was 12.47 in the left main pulmonary artery. On the basis of these results, primary malignancy of the artery or tumor embolus was suspected. Bronchoscopy to the nodular lesion in the lung was next performed. Because the nodule was located adjacent to the mediastinum, it was difficult to obtain material, and the biopsy result was negative. EBUS was then performed, which revealed that the masslike lesions were located inside the PAs and totally occluded the left lower lobe PA. EBUS-TBNA was performed to the mass in the left lower lobe PA (Fig 3). No intraoperative or postoperative complications occurred. The biopsy results showed spindle and polygonal cells with hyperchromatic nuclei arranged in vague fascicle without epithelioid arrangement (Fig 4). Immunostaining results were positive for smooth muscle antigen, vimentin, CAM5.2, and AE1/3; partially positive for CD31 and CD34; and negative for epithelial membrane antigen. According to the pathologic results, intimal sarcoma or sarcomatoid carcinoma was considered (Fig 4). On the basis of the pathologic and PET-CT results together, pulmonary sarcoma was diagnosed. Chemotherapy was not administered because the patient’s general condition deteriorated. He died 2 months after diagnosis.

Comment

PAS is a rare tumor, usually grows insidiously, and is symptom free until distant tumor embolism or severe pulmonary artery occlusion occurs. The pulmonary trunk is the most frequent location of PAS, followed by right PA, left PA, pulmonary valve, and right ventricular outflow [2]. This tumor grows regionally, with a tendency to metastasize primarily to the lung and mediastinal lymph nodes [2]. In the present case, CT yielded a differential diagnosis including pulmonary thromboembolism, tumor embolus, and primary malignancy of the artery. The FDG PET-CT results suggested malignant disease. However, it is often difficult to differentiate between PAS and pulmonary embolism by FDG uptake value [3]. Tumor emboli are usually microscopic, and patients typically present with dyspnea. In rare instances, the emboli can be macroscopic; in such cases, they are usually caused by tumors that have access to a venous plexus, such as hepatocellular, breast, and renal cell carcinomas [4]. In the present case, lung cancer was possible. However, no reports of lung cancer with macroscopic tumor embolus inside the main PAs and trunk have been published, to our knowledge. In the present case, tissue obtained from...
the intra-PA lesion by EBUS-TBNA showed sarcomatous tumor with positive epithelial marker immunostaining. Several reports exist regarding PAS with positive epithelial markers such as CAM5.2 and AE1/3 [5, 6]. PAS may show a wide variety of histologic differentiation: leiomyosarcomas, angiosarcomas, malignant fibrous histiocytomas, osteosarcomas, and others. Undifferentiated luminal sarcomas that do not neatly fall into one of these classifications are termed intimal. In consideration of the PET-CT and pathologic results together, PAS was diagnosed.

Only a few cases of PAS diagnosed neither by autopsy nor by surgical procedure have been reported. Penel and associates [7] reported a case of PAS with lung metastasis that was diagnosed by transbronchial lung biopsy (TBLB) [7]. Coli and associates [8] reported a case of PAS that was diagnosed by transvenous catheter biopsy. In the present case, EBUS with the use of real-time ultrasound images revealed that the mass lesions occluded the left lower PA, where no blood flow was observed. EBUS-TBNA was performed at that site, and tissue material was obtained. Park and colleagues [5] reported PAS diagnosed by EBUS-TBNA. These investigators obtained sufficient core tissue from the left pulmonary artery without

Fig 1. (A) Coronal view of an enhanced computed tomographic scan of the chest, showing filling defects in the bilateral main pulmonary arteries and a mass lesion of the right upper lobe of the lung. (B) Coronal view of an enhanced computed tomographic scan of the chest, showing a filling defect in the left pulmonary artery (arrowheads).

Fig 2. (A) Axial view of an enhanced computed tomographic scan of the chest, showing filling defects in the bilateral pulmonary arteries. (B) Axial view of a positron emission tomographic—computed tomographic scan of the chest, showing fluorodeoxyglucose uptake corresponding to the filling defects in the bilateral pulmonary arteries.

Fig 3. Endobronchial ultrasound image of the left pulmonary artery (PA), which is occupied by tumor; the needle is lying within the left pulmonary artery (white arrow).
complications, similar to the present case. Like TBLB, EBUS-TBNA carries the risk of bleeding in patients with pulmonary hypertension. However, EBUS has an advantage of being able to show blood flow with Doppler ultrasound in addition to showing real-time images. In this case, EBUS-TBNA was performed safely at the pulmonary artery site with no blood flow. EBUS-TBNA can be useful for examining intrapulmonary artery lesions and for the diagnosis of PAS.

References


Fig 4. (A) Tissue specimen obtained by endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) from the left pulmonary artery revealing malignant cells. (Hematoxylin and eosin; original magnification ×400). (B) Tissue specimen obtained by EBUS-TBNA from the left pulmonary artery revealing malignant cells immunostained with smooth muscle antigen (original magnification ×400).