Pulmonary Artery Sarcoma Diagnosed Using an Endovascular Catheter Forceps Biopsy

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Abstract:
We herein report a case of pulmonary artery sarcoma (PAS) in a 64-year-old woman. She was admitted to our hospital because of massive genital bleeding from endometrial cancer. Contrast-enhanced computed tomography (CT) revealed a left pulmonary artery mass and deep vein thrombosis. She underwent anticoagulant therapy for one year. However, the mass lesion gradually expanded. 18F-Fluorodeoxyglucose (FDG) positron emission tomography (PET)/CT showed a positive uptake of FDG by the mass. An endovascular catheter biopsy was performed for the differentiation of endometrial cancer metastasis or primary sarcoma. The biopsy specimen tissue comprised spindle-shaped cells. Thus, the patient was diagnosed with PAS.

Key words: pulmonary artery sarcoma, endovascular catheter forceps biopsy

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Introduction
Pulmonary artery (PA) sarcoma (PAS) is a rare malignant tumor originating from the PA. The clinical symptoms and imaging findings of PAS are similar to those of pulmonary thromboembolism and often lead to a misdiagnosis and subsequent inappropriate treatment. An endovascular catheter biopsy has been used to make a preoperative diagnosis (1); however, few reports discuss the details of the method.

We herein report a patient diagnosed with PAS based on the findings of an endovascular catheter forceps biopsy.

Case Report
A 64-year-old woman was admitted to our hospital because of massive genital bleeding due to endometrial cancer. Contrast-enhanced computed tomography (CT) on admission revealed a left PA mass (Fig. 1A) and deep vein thrombosis (DVT). The DVT was localized in the left superficial femoral vein and the bilateral below-the-knee deep vein (Fig. 1B, C). At that time, the PA mass was thought to be pulmonary thromboembolism from DVT. Hysterectomy and chemotherapy were chosen to treat endometrial cancer, and inferior vena cava (IVC) filter placement and anticoagulant therapy were chosen to treat the DVT. Within nine months of beginning anticoagulant therapy, the DVT had disappeared completely. However, contrast-enhanced CT revealed enlargement of the PA mass (Fig. 1D). Her shortness of breath had also increased gradually. She visited our department for an examination of the left PA mass.

On a physical examination, her blood pressure was 134/82 mmHg, and her pulse rate was 68 beats/min and regular. A chest examination revealed a Levine II/VI ejection systolic murmur at the left second intercostal space. No leg edema was present. Laboratory findings were within the normal range (D-dimer, 0.5 μg/mL; brain natriuretic peptide, 12.7 pg/mL). The arterial blood gas analysis revealed a partial pressure of oxygen of 83.9 Torr, PaCO2 of 38.1 Torr, and oxygen saturation of 97.6% (in room air). Chest X-ray imaging and electrocardiography findings were normal. 18F-Fluorodeoxyglucose positron emission tomography (FDG-PET)/CT showed a positive uptake of FDG by the PA mass (Fig. 1E). The uptake of FDG was not detected anywhere
catheters (Hyperion JR 4.0; ASAHI INTECC, Seto, Japan) showed complete occlusion of the left PA. Two 7-Fr guiding catheters (Hyperion JR 4.0; ASAHI INTECC, Seto, Japan) were inserted into the right ventricle and the distal right PA. Catheter measurements of the right ventricle and distal right PA pressure were 90/EDP 6 mmHg and 30/5 (16) mmHg, respectively. There was a pressure gradient between the distal right PA and the right ventricle, suggesting pulmonary artery stenosis due to the PA mass. An intravascular ultrasound examination (IVUS) (Eagle Eye Platinum; Philips, Amsterdam, The Netherlands) study revealed that the right PA lumen where the tumor was located was crescent-shaped (Fig. 2). However, the technique could not confirm the mass in the ostium of the left PA. Because of these limitations in the IVUS field size, an IVUS-guided biopsy was abandoned.

The endovascular biopsy was performed under angiographic guidance. For the three-dimensional CT image (Fig. 3B, D) and PA angiography (Fig. 3A, C), biopsy forceps were advanced through the 7-Fr guide-catheter to the PA trunk. Using 5-Fr biopsy forceps (Technowood, Tokyo, Japan), a biopsy of the PA mass occupying the left PA ostium was then performed (Fig. 4A, B). A rapid pathological diagnosis confirmed the presence of spindle-shaped cells, findings that differed from those of adenocarcinoma. A total of seven samples underwent a biopsy. After the procedure, PAG confirmed the absence of vascular injury. The total ra...
Radiation dose was 603 mGy, and the total contrast volume was 121 mL. The procedure time was 2 hours and 33 minutes, including the time to confirm the rapid pathological diagnosis.

The biopsy specimens showed that the tissue was composed of spindle-shaped or polygonal cells (Fig. 5). There was no epithelial structure. On immunohistochemical staining, the biopsy specimens were partially positive for smooth muscle actin but negative for CD31, CD34, and leukocyte common antigen. Based on these findings, the PA tumor was diagnosed as PAS. Surgical resection of PAS was chosen after a multidisciplinary cancer board conference.

The left lung and PA were resected. The tumor was approximately 5 cm in size, primarily in the PA, and extended to the left lung parenchyma and left hilar lymph node. The surgical pathological findings were similar to the biopsy results. The tumor surface was slightly sparse with respect to cellular components, and its substrate was fibrous tissue containing a hyaline-like component. Immunohistochemical staining of the pathology specimens revealed partial staining for smooth muscle actin and HHF35, a marker of muscle actin. The samples were negative for Cadesmon, Desmin, MyoD1, S-100, CD34, CD31. The mass was considered to be pulmonary artery intimal sarcoma of undifferentiated myofibroblastic origin in which specific differentiation could not be confirmed. The echocardiogram showed improvement of the right ventricle dilation, and the TR-PG level was 28.2 mmHg. These findings indicated the improvement of pulmonary artery stenosis.

Seven months after the operation, she was discharged and transferred for rehabilitation because of disuse syndrome due to operative stress.

Figure 2. An intravascular ultrasonography (IVUS) study revealed that the right PA lumen where the tumor was located was crescent-shaped. PA: pulmonary artery

Figure 3. Various imaging findings. (A and C) Angiographic examinations reveal a filling defect of the right main pulmonary artery. (B and D) Three-dimensional computed tomography (CT) images show a positional relationship between the pulmonary artery and the mass (pulmonary artery is blue, mass is green). (A and B) The anteroposterior view. (C and D) Left anterior oblique view at 60°.
Discussion

PAS is a rare malignant tumor with a poor prognosis. The first case was reported by an autopsy in 1923 (2). This lesion originates primarily from the main trunk of the PA, generally growing into the lumen and progressing along the blood vessels. The imaging findings of PAS are similar to those of pulmonary thromboembolism, and approximately half of cases are misdiagnosed as a pulmonary embolism (3). The diagnosis is often made at an autopsy (61% of cases) or post-surgery (4). A CT-guided needle biopsy (5) and endobronchial ultrasound-guided transbronchial needle aspiration (6) have been used to make the preoperative diagnosis. However, these procedures are difficult to perform when the tumor is localized in the PA.

A histological examination is important for the diagnosis of PAS. It is necessary to exclude metastasis cancer, and PAS must be differentiated from angiosarcoma, rhabdomyosarcoma and myxoma, which occur in a heart and a large vessel. In this case, she had a history of endometrial cancer and DVT. The histological examination was therefore very important for excluding metastasis cancer and chronic pulmonary embolism.

PAS is classified into two types: intimal and intramural. Intimal sarcoma usually shows fibroblast or myofibroblast differentiation. Histological findings of intimal sarcoma show proliferation of spindle-shaped cells in a myxoid background, alternating with a low-cellular collagen region. Because of the presence of fibroblasts, intimal sarcoma shows the diffuse expression of vimentin and may also express smooth muscle actin. In a tumor showing differentiation from vascular cells, CD31, CD34, and factor VIII may be expressed as endothelial markers (7). In our case, histological findings revealed spindle-shaped cell proliferation and positivity for smooth muscle actin, while other markers, such as CD31, CD34, and desmin, were negative. Therefore, she was diagnosed with pulmonary artery intimal sarcoma in which specific differentiation could not be confirmed.

An endovascular catheter biopsy is a reasonable method for diagnosing PAS that has developed into the PA. The first case of an endovascular catheter biopsy was reported in 1996 (1). Eight PAS cases that were diagnosed with an endovascular catheter forceps biopsy have recently been reported (8). An aspiration biopsy is less invasive and simpler to perform than a forceps biopsy; however, obtaining a sufficient sample for a biopsy can be difficult, such as when the tumor is hard (9). In the present patient, the surface of the mass was fibrous and hard. An aspiration biopsy was thus considered to be difficult, and only blood cell components were collected via an aspiration biopsy. Therefore, a forceps biopsy was required.

In our patient, we first attempted an IVUS-guided approach to confirm the mass during the biopsy. However, a biopsy under IVUS-guidance was difficult to perform in the PA because of the limited IVUS field size. Since an intracardiac echocardiography (ICE) provides a wider field of view than IVUS, we suggest using an ICE for confirming a PA mass during a biopsy. In this patient, a forceps biopsy...
was performed with angiographic guidance. The contrast-enhanced CT image was reconstructed three-dimensionally in advance; the positional relationship between the tumor and surrounding organs was thus clearly identified.

The biopsy was performed by confirming the position of the forceps from multiple directions. In a previous report (8), only a thrombus was obtained through a forceps biopsy in PAS patients. A forceps biopsy may miss a tumor, and the thrombus attached around a PAS may be removed. Therefore, a rapid pathological diagnosis was performed to confirm that a sufficient tissue specimen had been obtained from our patient.

Although the catheter forceps biopsy was successful in this patient, whether or not this approach can be safely performed in all cases is unclear. The catheter forceps biopsy is accompanied by risks such as pulmonary artery injury. In this case, the tumor was present in a protruding form in the left PA ostium, and the three-dimensional relationship could be observed well with CT. These findings meant that the catheter forceps could easily reach the PA tumor. It is necessary to discuss this approach on a case-by-case, especially concerning the localization of the PA tumor.

In our patient, a pulmonary artery forceps biopsy allowed the preoperative diagnosis of PAS without any complications. An endovascular forceps biopsy is a useful method for diagnosing a PAS located in the PA.

The authors state that they have no Conflict of Interest (COI).

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