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How to Do It

Endovascular Catheter Biopsy for the Diagnosis of Pulmonary Artery Sarcoma

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The diagnosis of pulmonary artery sarcoma (PAS) is challenging, and its definitive diagnosis is mainly confirmed using specimens obtained during surgery or autopsy. Endovascular catheter biopsy was performed in five patients with suspected PAS to establish a definitive diagnosis. Aspiration biopsy was performed in all patients, and forceps biopsy was performed in one patient. Three patients were diagnosed with PAS, and no definitive diagnosis was obtained in two patients with squamous cell lung carcinoma with pulmonary artery infiltration. Endovascular catheter biopsy is helpful in the diagnosis of PAS and should be performed when a tumor is suspected.

Keywords: pulmonary artery sarcoma, endovascular biopsy, preoperative diagnosis

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Introduction

Pulmonary artery sarcoma (PAS) is a rare and aggressive malignant tumor originating from the intimal layer of the pulmonary artery.[1)] The prognosis of PAS is extremely poor and requires early diagnosis followed by radical surgical resection to optimize survival time.[2)] However, the clinical symptoms and imaging findings of PAS are similar to those of pulmonary thromboembolism (PTE), which frequently leads to misdiagnoses and subsequent inappropriate treatment.[2)]

A definitive diagnosis of PAS is mainly confirmed using specimens obtained during surgery, autopsy, or, less often, during preoperative biopsy.[3–7)] Herein, we report on patients diagnosed with PAS via endovascular catheter biopsy.

Case Report

Patients and methods

This case series includes five patients who underwent endovascular catheter biopsy between April 2010 and September 2017. Based on clinical symptoms and the findings of contrast-enhanced chest computed tomography (CT) scans, PAS was suspected in all patients. Aspiration biopsy was performed in all five patients, and forceps biopsy was additionally performed in one patient. A comprehensive informed consent for the use of patient data was obtained from the patients prior to the procedure.

Procedures

A 5-F 11-cm sheath was inserted into the right internal jugular vein or right femoral vein, and a 5-F pigtail catheter (Terumo, Tokyo, Japan) or 4-F SOS catheter (Gadelius Medical, Tokyo, Japan) was advanced to the main pulmonary artery through the sheath using a 0.035-inch hydrophilic guidewire (SURF, Piolax, Yokohama, Japan). The right internal jugular vein approach was the first-choice method because a shorter catheter can reach the lesion and it is easier to perform suction. The femoral vein approach was selected in two cases because bronchial arteri-

Table 1 Characteristics of the patients and lesions

Case	Age (years)/ sex	Symptoms	Location of the tumor	Pathological diagnosis with endovascular catheter biopsy	Final diagnosis	Treatment	Outcome/follow-up
1	79/M	Loss of consciousness	PA, trunk, BPA	Sarcoma	PAS	Palliative care	Deceased/1 week
2	35/M	Chest pain, hemoptysis	RPA	Intimal sarcoma	PAS	Surgery, chemotherapy	Deceased/26 months
3	74/F	Hemoptysis	LPA with lung involvement	Not diagnosed	SCC with TBLB	Chemoradiotherapy	Alive/26 months
4	76/F	Palpitation, dyspnea	RPA	Intimal sarcoma with forceps biopsy	PAS	Chemotherapy	Alive/6 months
5	81/F	Dyspnea	PA, trunk, BPA	Not diagnosed	SCC with TBLB	Chemoradiotherapy	Alive/5 months

PA: pulmonary artery; BPA: bilateral pulmonary artery; RPA: right pulmonary artery; LPA: light pulmonary artery; PAS: pulmonary artery sarcoma; TBLB: transbronchial lung biopsy; SCC: squamous cell carcinoma of the lung

ography was performed simultaneously with the femoral arterial approach. Pulmonary angiography was performed in the main pulmonary artery trunk and in the right and left pulmonary arteries. The sheath was exchanged for a 7-F 90-cm guiding sheath (Shuttle sheath, Cook Medical, Bloomington, IN, USA) with a 0.035-inch J-type hydrophobic guidewire (Cook Medical, Bloomington, IN, USA), and a 7-F 100-cm VBL catheter (Cordis, Fremont, CA, USA) was inserted through the sheath into the diseased pulmonary artery. The catheter was then led to the filling defect using the preceding pulmonary angiogram. Vacuum suction was performed using a 20-mL syringe (BD, Franklin Lakes, NJ, USA) connected to the 7-F catheter. Suction was repeated three or more times until a sufficient amount of specimen was obtained. A 5.5-F endomyocardial biopsy forceps (Cordis, Fremont, CA, USA) was inserted through the 7-F catheter to clamp the tissue in the filling defect in one patient because the tumor was extremely hard, and it was difficult to obtain the specimen via suction. At the end of the procedure, a second pulmonary angiography was performed to confirm the absence of vascular injury. Systemic heparinization was not performed during the procedure, as bleeding can be fatal in pulmonary artery biopsy. Hematoxylin–eosin and immunohistochemical staining of the specimens were performed for histopathological evaluation.

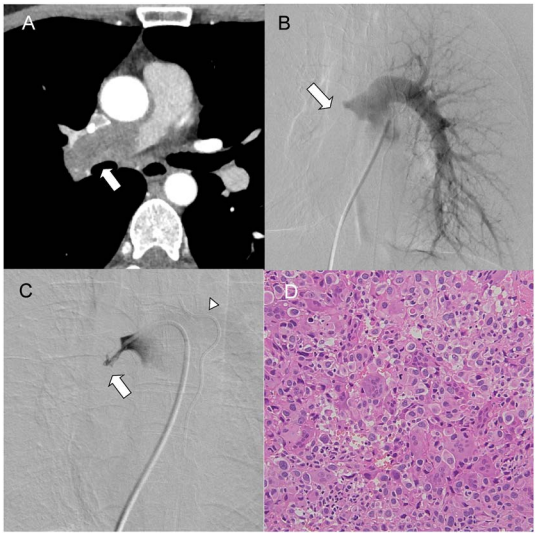

Fig. 1 Pulmonary artery sarcoma was suspected in a 35-year-old man who complained of chest pain and hemoptysis for several weeks.
(**A**) Contrast-enhanced computed tomography scan of the lung revealed occlusion of the right main pulmonary artery by a large mass-like filling defect. Endovascular catheter aspiration biopsy was performed. (**B**) Pulmonary angiography revealed complete occlusion of the right main pulmonary artery with abrupt disruption (arrow) and total perfusion defect of the right pulmonary parenchyma. (**C**) The catheter was connected to the filling defect of the right main pulmonary artery (arrow). Vacuum suction was performed several times by careful manipulation. Another catheter was cannulated into the right bronchial artery (arrowhead). (**D**) Histological examination revealed atypical pleomorphic cells with nuclear pleomorphism and some multinucleated cells (hematoxylin–eosin staining, ×200).

Results

Two of the five patients were male and three were female, with a combined mean age of 69.0 (range: 35–81) years (**Table 1**). The patients exhibited the following clinical symptoms: loss of consciousness, chest pain, hemoptysis, palpitation, and dyspnea. Contrast-enhanced chest CT scans revealed a thrombus-like mass occluding the central pulmonary artery in all five patients (**Fig. 1A**).

Endovascular catheter biopsy was successfully performed in all patients without any complications (**Figs. 1B** and **1C**). In two patients, aspiration biopsy specimens with hematoxylin–eosin staining revealed atypical pleomorphic cells as well as some multinucleated cells, and

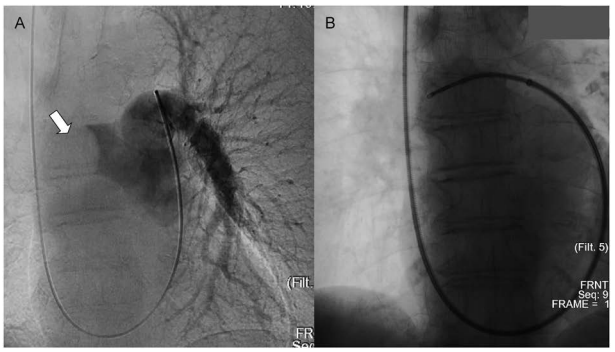

Fig. 2 Pulmonary artery sarcoma was suspected in a 76-year-old woman who complained of palpitation and dyspnea.
(**A**) Pulmonary angiography revealed complete occlusion of the right main pulmonary artery (arrow). (**B**) Vacuum suction was performed several times through a catheter inserted into the right main pulmonary artery. However, the tumor was extremely hard to suction a sufficient amount of specimen. We inserted a 5.5-F endomyocardial biopsy forceps and clamped the tissue.

the cell block preparation with immunohistochemical staining was diagnosed as PAS (**Fig. 1D**). In two patients, a definitive diagnosis could not be established using aspiration specimens due to an insufficient number of cellular components, and transbronchial lung biopsies in these patients revealed squamous cell lung carcinoma. The use of aspiration specimens did not obtain a definitive diagnosis in the last patient. However, a forceps biopsy was performed, and the patients were diagnosed with PAS (**Figs. 2A** and **2B**).

Among the three patients diagnosed with PAS, one died after a week of palliative care, one survived for 26 months with surgery and chemotherapy, and one is still alive and received chemotherapy for 5 months.

Discussion

Despite the recent advances in imaging modalities, the diagnosis of PAS remains challenging because its clinical symptoms and imaging findings are similar to those of PTE, which often leads to misdiagnosis and inappropriate treatment.[2] The present study aimed to establish a method for a definitive preoperative diagnosis of PAS with endovascular catheter biopsy. The histopathological diagnosis of PAS was confirmed in all three patients whose specimens were obtained during endovascular catheter biopsy. In the remaining two patients whose final diagnosis was squamous cell lung carcinoma with pulmonary artery infiltration, a definitive diagnosis was not established using endovascular catheter biopsy. Because endovascular catheter biopsy can directly approach a lesion in the vascular lumen, it is a reasonable diagnostic method for patients in whom PAS originates from the vascular intima and extends into the lumen. Conversely, our results suggest that endovascular catherer biopsy is less suitable for the diagnosis of lung carcinoma because fewer tumor cells are present in the vascular lumen at the infiltrated site.

In the present study, two patients were diagnosed by analyzing aspiration specimens using the cell block technique, which employs the retrieval of small tissue fragments and improves diagnostic accuracy.[8] Even if small specimens are collected during aspiration biopsy, it is possible to confirm the histopathological diagnosis of PAS using the cell block technique. We suggest that aspiration biopsy should be performed first, as it is less invasive and can be easily executed compared with other methods. However, the use of aspiration specimens may be insufficient for determining a definitive diagnosis when the tumor is extremely hard and suction cannot be appropriately performed. Even in such a case, it may be possible to obtain a sufficient amount of specimen with the endovascular approach by performing forceps biopsy. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) has also resulted in the successful diagnoses of PAS[9,10] due to its advantage of showing real-time tumor images. However, EBUS-TBNA poses a risk of bleeding in patients with pulmonary hypertension, thereby leading to massive hemoptysis.

Surgical resection followed by chemotherapy is the mainstay of PAS treatment. The prognosis without surgical resection is extremely poor. The reported median survival time and 5-year survival rate of patients are 11 ± 3 months and 0% without resection and 36.5 ± 20.2 months and 49.2% with resection, respectively.[2] In our study, one patient with PAS qualified for surgical resection after biopsy. However, two patients were not indicated for surgery because the tumors were widespread, and a surgical procedure was no longer considered advantageous. The less invasive biopsy technique described in the present study may aid the development of an appropriate therapy for such patients in the progressive stages of the disease.

Our study has a few limitations. First, the sample size is small owing to the rarity of the disease. Second, although we believe that it is best to perform aspiration biopsy first, it is difficult to judge whether a sufficient amount of specimen can be collected from each patient during the procedure. In combination with the procedure, bedside cytology may be necessary to ensure that a sufficient amount of specimen is collected. Despite these limitations, we successfully diagnosed PAS in all three patients using endovascular catheter biopsy without complications due to the intimal nature of the disease.

In conclusion, endovascular catheter biopsy is a safe and helpful technique in confirming the diagnosis of PAS, and it should be attempted promptly when a tumor is suspected.

Disclosure Statement

There is no conflict of interest for all authors.

Author Contributions

Study conception: YF, JK
Data collection: YF
Analysis: YF, JK
Investigation: YF, JK, TH, TS, CI
Writing: YF
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

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