Short Communication

Endovascular Catheter-guided Forceps Biopsy for the Diagnosis of Suspected Pulmonary Artery Sarcoma: A Preliminary Study of Eight Cases

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Introduction

Pulmonary artery sarcoma (PAS) is a rare malignant tumor that originates from the pulmonary artery (PA) with a poor prognosis.[1] Early diagnosis and radical surgical resection offer the only chance for survival.[2,3] As most PA sarcomas involve the PA trunk, making a preoperative histopathological diagnosis is quite difficult. So far, most PAS cases were reported with diagnosis made either at autopsy or intraoperatively with frozen sections.[4,5] Therefore, it will be very helpful if PAS can be diagnosed before surgery. For this purpose, some authors have attempted transcatheter suction biopsy to diagnose PAS preoperatively.[6,7] However, transcatheter suction biopsy often misses out the tumor as it does not provide sufficient core tissue. In our clinical practice and other reports, few of them succeeded.[8] In this preliminary study, a new technique of endovascular catheter-guided forceps biopsy (CGFB) was used to diagnose PAS. We describe the procedure and report results on a series of eight cases.

Methods

The Ethics Committee of Beijing Chao-Yang Hospital approved this study. Written informed consent was obtained from each patient before each invasive procedure. Between January 2012 and May 2015, 16 consecutive patients suspected with PAS were admitted in Beijing Chao-Yang Hospital for further diagnosis. Eight of the patients agreed to perform CGFB after right-heart catheterization. Details of the CGFB were described as followings: (1) A 6-F guiding catheter JR4.0 (Medtronic, Inc. Minneapolis, USA) was advanced to the PA with the help of wire. (2) The wire was removed, and the catheter was put in close touch with angiographic "thrombus-like" substance. (3) Endomyocardial biopsy forceps (Argon Medical Devices, Inc. Plano, USA) were inserted directly in touch with the mass, once resistance was sensed, clamp the tissue quickly and then pulled out the forceps. The operation was repeated until enough tissues were obtained [Figure 1]. Hematoxylin-eosin staining and immunohistochemical staining of the specimen were carried out for evaluation.

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Results

Of the eight patients agreed for CGFB, three were men and five were women. The mean age of the patients was 52.4 years (range, 32–75 years), [Table 1]. Of eight patients, six were initially diagnosed as pulmonary embolism and treated with anticoagulants (6/6) or thrombolysis (2/6) without any improvement. PAS was suspected at first visit in two patients. The thrombus-like mass occluded the central pulmonary arteries in all of the eight patients. Three patients occluded the right PA (RPA), two in the left PA, and two

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located both in the PA trunk and RPA, and in one case, the mass involved the outflow tract of right ventricle, PA valve, PA trunk, and both sides of the pulmonary arteries. Lung involvement was also identified in three patients [Figure 2].

All the eight patients were performed CGFB successfully and tissue samples were gained for histological examination. PAS was confirmed in five patients, and chronic thromboembolic pulmonary hypertension (CTEPH) was considered the correct diagnosis with the histological findings from CGFB and confirmed after pulmonary thromboendarterectomy later [Figure 3]. While in two patients, there were only necrotic tissue and thrombus detected from the harvested sample, one of the patients was then confirmed PAS with computed tomography (CT)-guided percutaneous lung biopsy 4 months later.

Of the five patients diagnosed with PAS after CGFB, two died between 3 and 4 months without any specific treatment. One was still alive at the time of the last follow-up (14 months) with chemotherapy. The other two was not received any treatment yet. Among the other three patients not confirmed as PAS after CGFB, one was diagnosed as CTEPH and improved dramatically after surgery as followed up for 4 months. One of the patients was then confirmed to have PAS with CT-guided percutaneous lung biopsy 4 months later. Moreover, the other one suffered from sudden cardiac death after 15 months without any specific treatment.

All the eight patients tolerated the procedure well. There were no complications such as bleeding and perforation during and after CGFB in eight patients.

Table 1 shows demographic and clinical characteristics of eight cases with suspected PAS.

Discussion

PAS is a rare and lethal neoplasm. It generally occludes the central pulmonary arteries and is frequently misdiagnosed

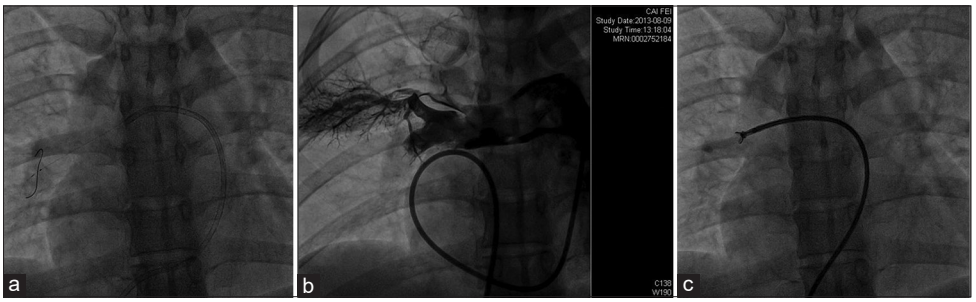

Figure 1: (a) The insertion of pulmonary artery catheter through a wire. (b) Selective pulmonary angiography of the right lung showed filling defect of the right main pulmonary artery and cutoff of the right superior and inferior pulmonary artery. (c) Catheter-guided forceps biopsy was performed for the lesion in the right inferior pulmonary artery.

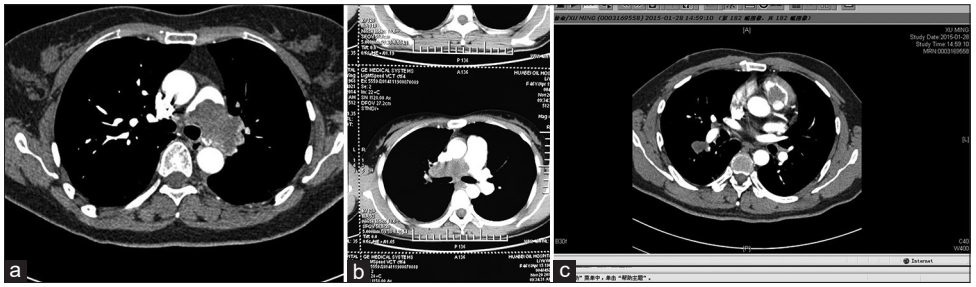

Figure 2: (a) Computed tomography scan showing a lobulated soft tissue mass filling the main and left pulmonary artery in one case with pulmonary artery sarcoma. (b) Filling defects in right pulmonary artery in another case with pulmonary artery sarcoma. (c) The lung and the outflow tract of right ventricle involvement in another case with Pulmonary artery sarcoma.

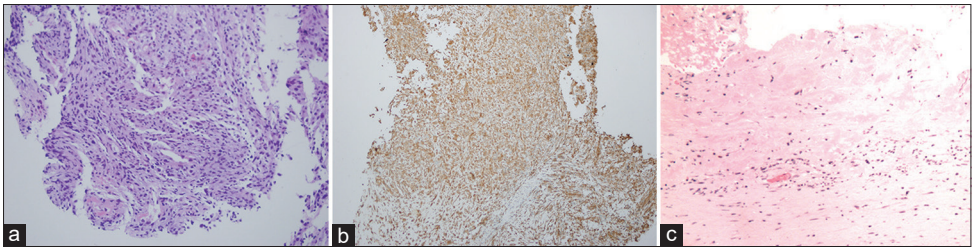

Figure 3: (a) The histological examination revealed poorly differentiated spindle-cell malignancy (H and E, original magnification ×200). (b) Tumor cells show definite immunoreactivity for vimentin (immunostaining, original magnification ×100). (c) Histological examination revealed fibrous and degenerative tissue (H and E, original magnification ×100) and was later confirmed as chronic thromboembolic pulmonary hypertension after surgery. H and E: Hematoxylin and Eosin.

Table 1: Demographic and clinical characteristics of eight cases with suspected PAS

Case number	Gender/ age (years)	Main complaints	Initial diagnosis	Site of tumor	Pathological diagnosis with CGFB sample	Final diagnosis	Treatment	Outcome and follow-up
1	Male/75	Dyspnea	PE	RPA	Necrotic tissue	–	NS	Died, 15 months
2	Female/49	Hemoptysis, chest pain, dyspnea	PE	LPA + lung involvement	Intimal sarcoma	PAS	Chemotherapy	Alive with disease, 14 months
3	Female/32	Dyspnea	PAS	RPA	Intimal sarcoma	PAS	NS	Died, 3 months
4	Male/59	Chest pain, dyspnea	PE	PA trunk + RPA	Intimal sarcoma	PAS	NS	Died, 4 months
5	Female/46	Chest pain, dyspnea	PE	RPA + lung involvement	Thrombus	PAS (diagnosed by CT-guided percutaneous lung biopsy)	NS	Alive with disease, 4 months
6	Male/47	Chest pain, dyspnea, hemoptysis	PE	LPA	Fibrous and degenerative tissue, consistent with CTEPH	CTEPH	PET	Alive with improvement, 6 months
7	Female/69	Chest pain and dyspnea	PE	Outflow tract of right ventricle + PA valve + PA trunk + RPA + LPA	Sarcoma	PAS	NS	Alive with disease 1 months
8	Female	Cough and dyspnea	PAS	PA trunk + RPA + lung involvement	Sarcoma	PAS	Chemotherapy	Alive with disease 2 weeks

CTEPH: Chronic thromboembolic pulmonary hypertension; PE:Pulmonary embolism; LPA: Left pulmonary artery; PA: Pulmonary artery; RPA: Right pulmonary artery; NS: Not specific, PET: Pulmonary thromboendarterectomy; PAS: Pulmonary artery sarcoma; CT: Computed tomography; CGFB: Catheter-guided forceps biopsy.

as acute or chronic pulmonary thromboembolism.[9-11] This misdiagnosis contributes to its poor prognosis as it delays making the correct diagnosis and administering the appropriate treatment. The definite diagnosis of PAS is based on pathological examination, and so far, the majority of specimens is taken by surgery or autopsy. It was reported that an early diagnosis of PAS may improve its prognosis.[12] Therefore, it will be very helpful if it can be diagnosed preoperatively. However, the diagnosing of PAS is still considered very hard before surgery or autopsy although many attempts have been made. Endovascular aspiration biopsy has been attempted to diagnose PAS, but with limited success.[6-8] Insufficient core tissue is the main concern. Endobronchial ultrasound-guided transbronchial needle aspiration has also been reported with a successful diagnosis of PAS. However, the increasing complications of bleeding have been questioned.[13-15]

In our cases, endovascular biopsy was performed through catheter-guided forceps clamp. Results showed that six of the eight patients get histopathological diagnosis with the samples obtained through CGFB, including one diagnosed as CTEPH and five as PAS. In PAS, the tumor tissue is often covered with *in situ* thrombus and necrotic tissues. In comparison with endovascular aspiration biopsy, CGFB may obtain more core tissues. In our study, of the eight cases, only necrotic tissue or thrombus was obtained through CGFB in two patients. As one of the two patients was later confirmed as PAS, the definite diagnosis was not available for the other one. There were probabilities that CGFB may miss out the tumor. In our experience, forceps should be inserted in touch with the mass and clamp the tissue when resistance was sensed as quickly as possible. The procedure may be repeated until the core tissues were obtained, which have more chance to get the real lesion site.

In one case, there was no distinct morphologic and immunohistologic differentiation detected; thus, the origin of the tumor was not clear through CGFB. Not enough tissue for histopathological analysis and poor differentiation of the tumor were possible reasons.

There might be another concern about the safety of CGFB; however, CGFB was safe in our cases. No complications occurred in all of the eight patients. CGFB is comparatively less invasive and easier to be carried out, and the materials used in CGFB are easily available in a catheter laboratory. However, it has to be admitted that the technique takes risk of bleeding, perforation, etc.

In conclusion, our cases suggest that CGFB is a safe and feasible approach to the tissue diagnosis of suspected PAS preoperatively. CGFB can be used as a diagnostic option for PAS according to the tumor location although additional experience is required.

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Conflicts of interest

There are no conflicts of interest.

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