Bronchial-pulmonary arterial fistula with primary racemose hemangioma

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Abstract
Bronchial-pulmonary arterial fistula (BPAF) is a rare vascular malformation complicated with racemose hemangioma. We report a case of a 65-year-old male with BPAF with primary racemose hemangioma. Bronchial arteriography demonstrated convolution, dilation, and aneurysm connected with pulmonary artery, suggesting the presence of BPAF, in the left upper lobe. Since a 20-mm sized aneurysm of bronchial artery and BPAF coexisted, he underwent ligation of bronchial arterial aneurysm and left upper lobectomy to prevent hemoptysis. As neither history of prior illness nor histopathologic findings of vascular inflammation was seen, the present case demonstrated BPAF with primary racemose hemangioma. Establishment of non-invasive treatment strategy for BPAF is urgently required.

Introduction
Bronchial-pulmonary arterial fistula (BPAF) is complicated with racemose hemangioma, a rare malformation of bronchial vasculature, and often causes life-threatening hemoptysis. Racemose hemangioma has been reported since the end of 1970s [1], and considered to be classified into primary and secondary forms, respectively, due to congenital malformation and broncho-pulmonary inflammation [2]. Bronchial arterial embolization (BAE) as the first-line therapy, bronchial arterial ligation, and surgical resection if necessary, are tools of therapeutic management for racemose hemangioma to prevent hemoptysis. We present a case report of a 65-year-old male with BPAF and primary racemose hemangioma.

Case Report
The patient was a 65-year-old male who experienced exertional dyspnea (Modified Medical Research Council [mMRC] grade 2) and palpitation in recent 3 months. A chest X-ray showed a nodular lesion in left upper field (Fig. 1A), and computed tomography (CT) revealed convolution and dilation of pulmonary vessels (Fig. 1B and C). Three-dimensional CT revealed convolution and aneurysm of bronchial artery (Fig. 1D). Bronchial arteriography demonstrated 4-mm dilated convolution and 20-mm sized pooling of contrast injection at segment 1 + 2 in the left upper lobe. Outflow from the vascular lesion into pulmonary artery of 1 + 2 was also present during the early arterial phase, suggesting the presence of fistula between bronchial and pulmonary artery, (i.e. BPAF) (Fig. 1E). Pulmonary arteriography revealed a decrease in filling of contrast injection into A 1 + 2 (Fig. 1F). The decrease in filling of contrast injection into pulmonary artery appeared to be explained by a possibility that higher blood pressure of the BPAF disturbed depiction of pulmonary arteriography. There was no fistulous tract between pulmonary artery and vein. Bronchoscopy was not performed because of the patient’s anxiety about airway bleeding.
Since 20-mm sized aneurysm of bronchial artery and BPAF coexisted, the patient underwent left video-associated thoracotomy, and then ligation of the bronchial arterial aneurysm and upper lobectomy were taken. Because the aberrant bronchial artery lesion reached to the lingular bronchus, it was hypothesized that upper lobectomy rather than upper division segmentectomy appeared to be the appropriate treatment. He was discharged home without any incident on postoperative day 5 and resolved his exertional dyspnea. No overt appearance of new aneurysm in bronchial artery was seen in follow-up CT 1 year later.

Pathological examination revealed irregular thickness of media and abrupt calcification in the vascular wall of dilated bronchial arteries. No findings of vasculitis, including inflammatory infiltration and plexiform lesion, were seen (Fig. 2).

There was no history of infection, foreign body ingestion, trauma, upper respiratory bleeding or travel, and no teleangietasia was seen on physical examination. There was no familial history of coagulopathy, vascular malformation, epistaxis, stroke, or respiratory illness.

**Figure 1.** (A) Chest X-ray shows nodular lesion (arrow) in left upper field. (B) Chest computed tomography (CT) shows a well-demarcated nodule adjacent to dilated pulmonary vessel in left segment of 1 + 2. (C) Contrast enhanced CT reveals convolution and dilatation of pulmonary vessels in left segment of 1 + 2 by (arrows). (D) Three-dimensional CT reveals convolution and aneurysm of bronchial artery (arrows). (E) Bronchial arteriography demonstrates 4-mm dilated convolution and 20-mm sized pooling of contrast injection at segment 1 + 2 in the left upper lobe (arrows). Outflow from the vascular lesion into pulmonary artery of 1 + 2 was also stained (dashed arrow). (F) Pulmonary arteriography reveals a decrease in filling of contrast injection in A 1 + 2 (arrow).

**Figure 2.** Surgical specimen shows irregular thickness of media (arrow) and abrupt calcification (dashed arrow) in vascular wall of dilated bronchial arteries. (Elastica van Gieson stain, x5).
Discussion

Racemose hemangioma is characterized by not only a dilated and convoluted bronchial artery but also a vascular hyperplasia causing abnormal anastomosis to adjacent vessels [3]. Racemose hemangioma creates a fistula to the pulmonary vasculature; however, the incidence of fistula from bronchial racemose hemangioma to pulmonary arteriovenous malformation (PAVM) has been estimated approximately at 4% among PAVM [4]. Narato et al. summarized 34 Japanese cases of bronchial racemose hemangioma [5]. The authors demonstrated that 30 cases caused hemoptysis and 25 cases possessed lesions in right lungs among 16 male and 18 female patients with a mean age of 49 years (range 19–80).

Therapeutic candidates for racemose hemangioma include BAE, bronchial arterial ligation, and surgical resection. However, the typical treatment has not been established. According to review by Narato et al. in Japan, whereas lobectomy was performed most often approximately at 38%, BAE was carried out at 29% for racemose hemangioma [5]. BAE appears to be non-invasive and recognized as the first line therapy for racemose hemangioma; however, it raises several concerns about the failure of embolus and the reanastomosis of vascular malformation after BAE treatment. Furthermore, it also raises a possibility of increased risk for erratic embolus, and bleeding in the presence of fistulous tract from racemose hemangioma to adjacent vessels, such as BPAF, after BAE treatment [4]. Because a 20-mm sized aneurysm of the bronchial artery and BPAF coexisted, BAE may fail, resulting from both inappropriate embolization and bleeding. Pulmonary arteriography revealed the decrease in blood flow at pulmonary artery adjacent to BPAF, indicating a disturbance of pulmonary blood flow due to higher blood pressure from BPAF. These findings suggested that although the patient was not in a critical condition, the malformation of BPAF was severe and suggested an increase in risk for hemoptysis in the future. It was hypothesized that ligation of the bronchial arterial aneurysm and left upper lobectomy rather than BAE appeared to be a suitable tool in the present case.

Racemose hemangioma has been classified into two types, congenital vascular malformation as a primary form and subsequent vascular malformation by broncho-pulmonary inflammation as a secondary form [2]. Several acquired process, such as pulmonary infection, chronic broncho-pulmonary inflammation, trauma, and neoplasm including lung cancer and lymphoma, may contribute to the development of secondary racemose hemangioma [4]. None of these acquired causes were seen in the present case. Furthermore, as pathological examination revealed no findings of vasculitis, the present case demonstrated BPAF with primary racemose hemangioma.

We report here a case of BPAF from primary racemose hemangioma. Since a 20-mm sized aneurysm of the bronchial artery and BPAF coexisted, the patient underwent ligation of bronchial arterial aneurysm and left upper lobectomy rather than BAE. Establishment of non-invasive treatment strategy for BPAF is urgently required.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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