—Case Reports—

Two Cases of Bronchial Artery Racemose Hemangioma Successfully Treated with Bronchial Artery Embolization

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Rupture of a racemose hemangioma causing dilatation and tortuosity of the bronchial artery can result in massive bleeding and respiratory failure. Bronchial artery embolization (BAE) can treat this life-threatening condition, as we show in two cases. The first case was of an 89-year-old female complaining of sudden-onset chest and back pain. Bronchial artery angiography demonstrated a racemose hemangioma with a 2 cm aneurysm. The second case was of a 50-year-old male with hemoptysis and dyspnea, eventually requiring intubation. Bronchial arteriography showed a racemose hemangioma and a bronchial artery-pulmonary arterial fistula. BAE was successfully performed in both cases, with no recurrent hemorrhage. Therapeutic interventions in bronchial artery racemose hemangiomas include lobectomy or segmentectomy, bronchial arterial ligation, and BAE. BAE should be considered as first-line therapy for bleeding racemose hemangiomas of the bronchial artery because of its low risk of adverse effects on respiratory status, minimal invasiveness, and faster patient recovery.
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Key words: cardiovascular abnormalities, bronchial arteries, radiology, interventional, embolization

Introduction

Bronchial artery racemose hemangioma is a rare vascular anomaly characterized by dilated and convoluted bronchial arteries. Patients complain of cough and dyspnea, which can progress to respiratory failure and hypovolemic shock. There have been a limited number of reports on bronchial artery racemose hemangioma and very few on bronchial artery embolization (BAE) in emergency situations. We describe two cases of acutely hemorrhagic bronchial artery racemose hemangioma successfully treated with urgent BAE.

Case Report

Case 1: An 89-year-old female was admitted with sudden-onset chest and back pain. She was tachypneic (RR 26/min) and hypoxemic (PaO_2 131 mmHg on 10 L/min oxygen). Contrast-enhanced chest computed tomography (CT) showed a mediastinal hematoma and a large right and smaller left hemothorax (**Fig. 1A, B**). Three-dimensional reconstruction of image data (**Fig. 1C**) revealed the right bronchial artery to be dilated and convoluted, with an aneurysm measuring 2 cm in greatest diameter. No extravasation of contrast was detected. After intubation, emergency bronchial artery angiography revealed a racemose hemangioma arising from the distal segment and the aneurysm arising from the proximal segment of the right bronchial artery (**Fig. 2A**).

We performed selective right bronchial artery angiography and occluded the aneurysm and its feeding vessel employing a previously reported[1] method using detachable coils and N-butyl-2-cyanoacrylate (NBCA) (**Fig. 2B**). Followup aortography revealed no collateral arterial supply to the distal branch (**Fig. 2C**). On the second day after admission, the patient was extubated. Her vital signs

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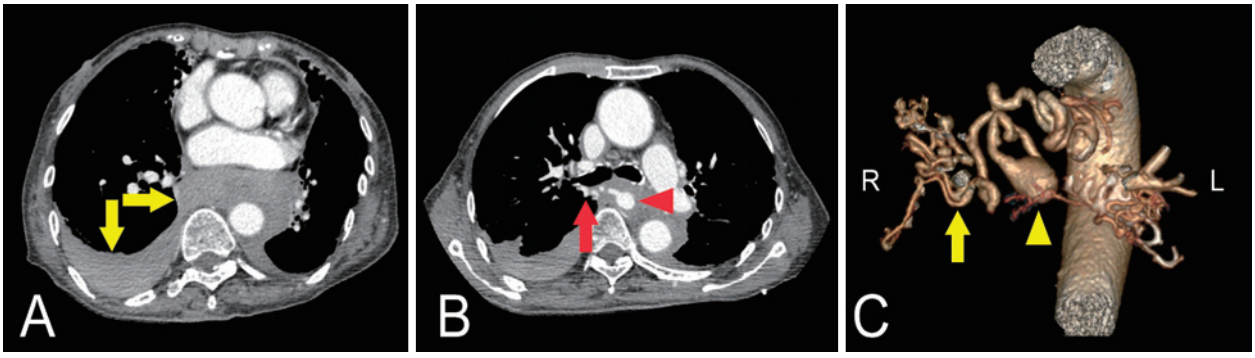

Fig. 1 Case 1. (A) Chest contrast-enhanced computed tomography (CT) scan showed mediastinal hematoma and pleural effusions (arrow). (B) Chest contrast-enhanced CT scan revealed a dilated and convoluted bronchial artery (arrow) and an area of focal dilatation (arrowhead). (C) 3D reconstruction of CT scan showed dilation and tortuosity of the bronchial artery (arrow) with a 1.2. × 1.5 × 2.0 cm aneurysm (arrowhead).

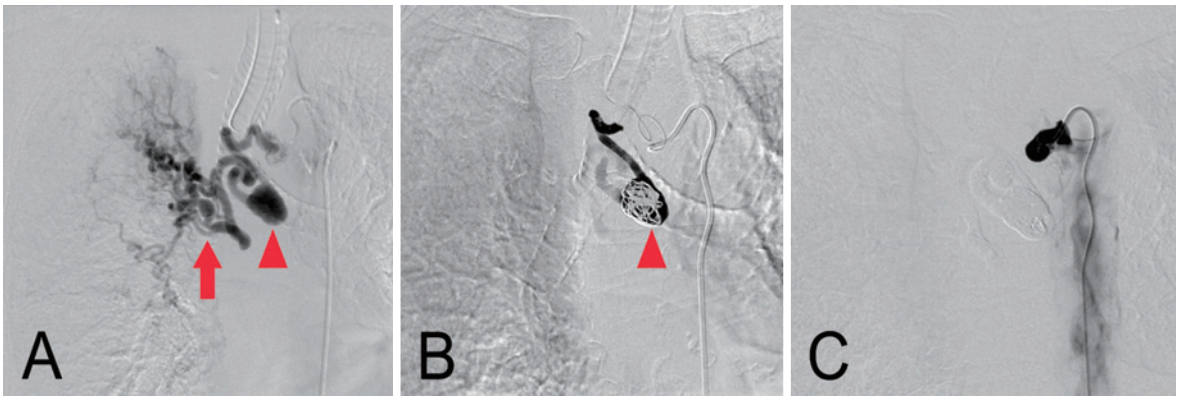

Fig. 2 (A) Bronchial artery angiogram demonstrated racemose hemangioma (arrow) and the aneurysm (arrowhead). (B) The aneurysm was embolized with detachable coils and N-butyl-2-cyanoacrylate (arrowhead). (C) No residual racemose hemangioma was detected by aortography following embolization of the aneurysm.

were stable, and she no longer needed supplemental oxygen by the sixth day. She was transferred to another hospital for rehabilitation.

Case 2: A 50-year-old man presented to his primary care doctor with a cough that had become increasingly productive for the past month, mild hemoptysis for one week, and dyspnea for 3 days. He was admitted to an outside hospital. Massive hemoptysis on the third day after admission occurred, and he was intubated. Chest CT scan showed consolidation in the right upper and left lower lobes (**Fig. 3A, B**). Bronchoscopy could not visualize a bleeding source due to hemorrhage. He was transferred to our hospital. Contrast-enhanced chest CT revealed a dilated and tortuous right bronchial artery without evidence of active extravasation (**Fig. 3C**). Bronchial artery angiography following the CT scan revealed ectasia and tortuosity of both upper and lower lobe branches of the right bronchial artery at the level of T4-6 (**Fig. 4A**) and a bronchial artery-pulmonary arterial fistula, visualized as a faint parenchymal blush (**Fig. 4B**). We performed BAE with gelatin sponge to occlude the two branches of the right bronchial artery. We did not use NBCA to avoid the permanent occlusion of the potential feeders of spinal cord[2]. After the procedure, marked reduction of blood flow was seen in the right bronchial artery (**Fig. 4C**). Following transfusion on the day of the procedure, there was no postprocedural hemoptysis and the hematocrit remained stable. He was extubated on the 12th day, and no longer required supplemental oxygen 24 days from the first admission to the outside hospital. Subsequent bronchoscopy performed during hospitalization detected a nodular lesion with a reddish bloody surface at the takeoff of the right lower lobe superior segmental bronchus (B6), corresponding to the tortuous vascularity, and indicative of a racemose hemangioma (**Fig. 5**). He was discharged home on the 28th day without any recurrence of hemoptysis.

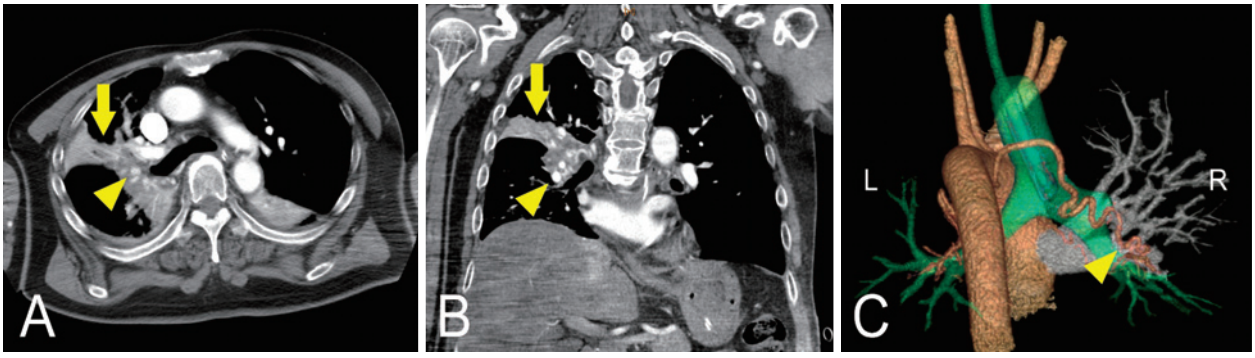

Fig. 3 Case 2. (A, B) Chest contrast-enhanced computed tomography (CT) scan showed consolidation in the right upper lobe (arrow), and a dilated and convoluted bronchial artery (arrowhead). (C) Posterior view of 3D reconstruction of a contrast-enhanced CT scan again reveals a dilated and convoluted bronchial artery (arrowhead).

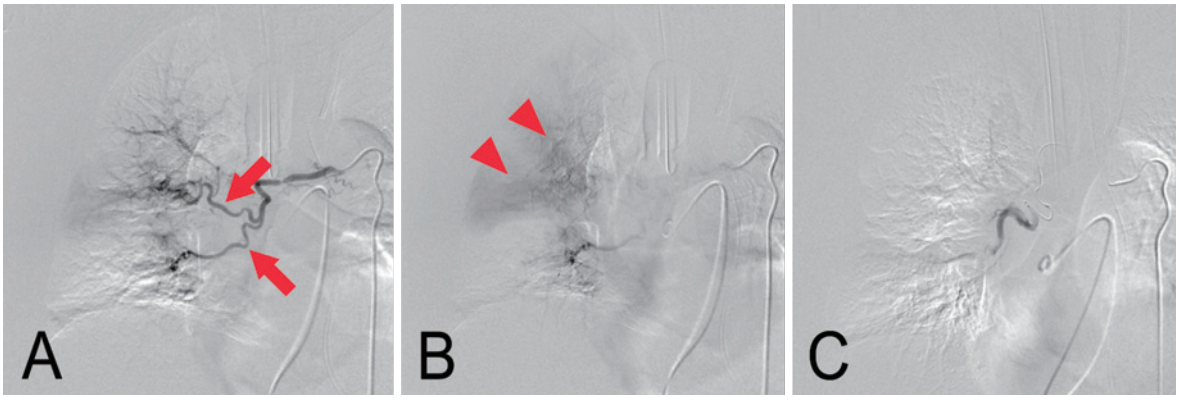

Fig. 4 (A) Early phase of bronchial artery angiogram demonstrated vascular supply from upper and lower lobe branches of the right bronchial artery (arrows). (B) The delayed phase of the bronchial artery angiogram demonstrated the presence of a bronchial artery-pulmonary arterial fistula as depicted by faint pulmonary parenchymal opacification (arrowheads). (C) After bronchial artery embolization with gelatin sponge, the dilated and convoluted bronchial artery and bronchial artery-pulmonary arterial fistula had disappeared. A small amount of contrast remained in the main bronchial artery.

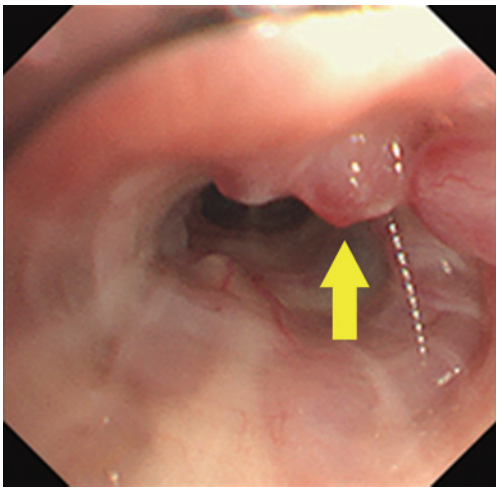

Fig. 5 Bronchoscopy performed on the 27th day showed a mass at the takeoff of the bronchus to the superior segment of the right lower lobe (B6) (arrow).

Discussion

Racemose hemangioma of the bronchial artery was first described in the 1970s[3]. It has been considered to be a vascular anomaly with a dilated and tortuous bronchial artery, causing symptoms such as cough, hemoptysis, and dyspnea[4]. In 2009, Narita et al. reported cases of bronchial artery racemose hemangioma in Japan across all age groups, with a mean age of 53 years. The male and female genders were equally affected. Mild to severe hemoptysis was the presenting symptom in 83%[5]. Racemose hemangioma of the bronchial artery is often diagnosed based on findings on CT scan and/or bronchial artery angiography. Gupta et al. presented the hypothesis that the bronchial artery may be abnormally hypertrophied when the diameter of the artery is ≥ 2 mm, and/or the course of it is visualized up to the pulmonary hilum on CT[6]. Racemose hemangioma of the bronchial artery is occasionally accompanied by aneurysms, as in Case 1[7]. In

addition, vascular hyperplasia, which leads to anastomosis or fistulization to adjacent vessels, such as the pulmonary artery or vein, often coexists, as in our cases[8,9]. The presence of a bronchial artery-pulmonary arterial fistula is a risk factor for massive hemoptysis since the bronchial artery, supplied by the systemic circulation, has higher pressure and velocity than those of the pulmonary artery[10,11]. For these reasons, bronchial artery racemose hemangiomas can be life-threatening, as in our cases.

Bronchial artery racemose hemangioma is classified into two types based on its development: primary and secondary. The primary type is a congenital abnormality, while the secondary type develops from chronic bronchial or pulmonary inflammatory disease such as pneumonia and tuberculosis, trauma, and malignant neoplasm, including lung cancer and lymphoma[10]. It is hypothesized that these lesions cause pulmonary arterial constriction or occlusion, which increases local demand for oxygen, thereby causing compensatory development of the bronchial artery[12]. Histologically, racemose hemangiomas have a thickened intima due to hyperplasia of elastic fibers and dilated lumens, accompanied by inflammatory changes in the surrounding tissues in secondary cases[5,13]. Bronchoscopy reveals subepithelial mass lesions, which were reported in 79% of cases Yokoyama et al. reviewed; and in 21% of those cases the lesions were pulsatile[14]. Again, in many cases, racemose hemangiomas of the bronchial artery fistulize to the pulmonary artery or vein, increasing the risk of life-threatening hemoptysis. The review by Yokoyama reported 64% of cases had bronchial artery-pulmonary arterial fistulas[14]. Case 1 was considered a primary case based on the absence of chronic inflammatory changes in her lung. Case 2 also appeared to be a primary case. The patient had developed respiratory failure due to the bronchial artery-pulmonary arterial fistula.

Definitive therapeutic management for bronchial artery racemose hemangioma has not been established. However, BAE should be considered as a first-line treatment because it is minimally invasive, takes a short amount of time, and can preserve lung function[10]. BAE was first described by Rémy in 1974[15] and has been performed for benign and malignant causes of hemoptysis. Based on the systematic review conducted by Panda, there was a clinical success rate of 70-99%[16]. Materials for BAE include gelatin sponge, coils, and NBCA. Gelatin sponge is absorbable within 2-6 weeks[6], while coils and NBCA are permanent. Gelatin sponge is helpful in the initial treatment to confirm the response to BAE, meanwhile, additional embolization using permanent materials may be required thereafter. After coil embolization, new arterial feeders may develop if coils are not placed sufficiently at the source of bleeding. NBCA is a liquid material which polymerizes quickly after injection; however, inadequate maneuver can cause an influx of NBCA into the pulmonary or systemic circulation[9]. Coils and NBCA can be used in combination, as in Case 1, to gain the firmer embolization effect[1]. Materials for BAE are chosen depending on morphology, anatomy, and the degree of hemorrhage. Other than BAE, surgical resection and bronchial arterial ligation are the other available therapies, though these are associated with higher morbidity and mortality rates[7]. Surgical resection can provide definitive treatment without risk of rebleeding even in cases complicated by bronchial artery-pulmonary arterial fistula and collateral circulation, but it is very invasive and decreases pulmonary function. Bronchial arterial ligation is sometimes selected to avoid loss of pulmonary function[17]. However, it cannot completely avoid rebleeding through the establishment of collateral circulation[17,18]. BAE also has some risks: difficulty in controlling bleeding, embolization of other arteries such as the pulmonary artery and anterior spinal artery, rebleeding and re-anastomosis after the procedure, and fistulization to the trachea[19–21]. However, we believe that BAE should be considered as a first-line treatment because of its minimally invasive nature, which makes it possible to treat a wide variety of patients, and because of its short recovery time, as seen in our cases.

Conclusion

We report two cases of bronchial artery racemose hemangiomas treated with BAE. BAE should be considered as first-line therapy for bleeding bronchial artery racemose hemangiomas because it has fewer adverse effects on respiratory status, is less invasive, and results in faster patient recovery.

Informed Consent: The patients provided written permission for publication.

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Conflict of Interest: No conflicts of interest declared.

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