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Case Report

A case of primary racemose hemangioma with endobronchial lesions demonstrating recurrent hemoptysis initially treated with bronchial arterial embolization

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ABSTRACT

Primary racemose hemangioma of the bronchial artery (RHBA) is one of the causes of massive hemoptysis. A 72-year-old woman was admitted to our hospital with recurrent hemoptysis. Bronchoscopy showed an endobronchial lesion, and the angiography of the right bronchial arteries indicated RHBA. Bronchial arterial embolization (BAE) was performed to prevent hemoptysis. Although the endobronchial lesion shrank after the first BAE, the lesion re-increased and caused massive hemoptysis. A thoracoscopic right upper lobectomy was performed, and hemoptysis did not recur. Therefore, in cases of RHBA where there is recurrent hemoptysis and the endobronchial lesions that remain after BAE, additional treatments should be considered.

1. Introduction

Primary racemose hemangioma of the bronchial artery (RHBA) is a rare abnormality of the bronchial artery that causes fatal hemoptysis [1,2]. Although bronchial arterial embolization (BAE) is minimally invasive and superior because embolization can be performed directly after bronchial angiography, thereby making BAE the first-choice treatment for RHBA with hemoptysis, recurrence is a common complication of BAE [3,4]. However, the overall immediate clinical success rate of BAE is high [5].

Although endobronchial lesions are often present in RHBA [2], the changes in the lesion after BAE are unclear. It is also unclear whether it is an indicator of recurrent hemoptysis.

Herein, we describe a case of primary RHBA with endobronchial lesions that had recurrent hemoptysis and was initially treated with BAE.

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2. Case report

A 72-year-old woman was admitted to our hospital for recurrent hemoptysis. Three weeks before admission, she had experienced hemoptysis and had been hospitalized in a nearby hospital. The patient's symptoms improved without specific treatment. However, she had recurrent hemoptysis and was admitted twice after the initial hospitalization. She was referred to our hospital for further treatment. Physical examination and laboratory tests, including a blood cell count and serum biochemical analysis, were performed upon admission to our hospital and showed normal results. However, chest radiography showed ground-glass attenuation (GGA) in the right middle and lower lung fields. Contrast-enhanced computed tomography (CT) also showed GGA in the right middle lobe and an endotracheal shadow in the right upper bronchial entrance (Fig. 1). There was no cluster of blood vessels in its shadow. Bronchoscopy results revealed an elevated lesion at the entrance of B3 (Fig. 2A). Although there was no pulsation in the lesion, massive bleeding appeared after a biopsy of the lesion with forceps (Movie 1). The patient's respiratory condition worsened due to massive bleeding in the bronchi; thus, intensive care with endotracheal intubation was temporarily administered. Bronchoscopy performed af-

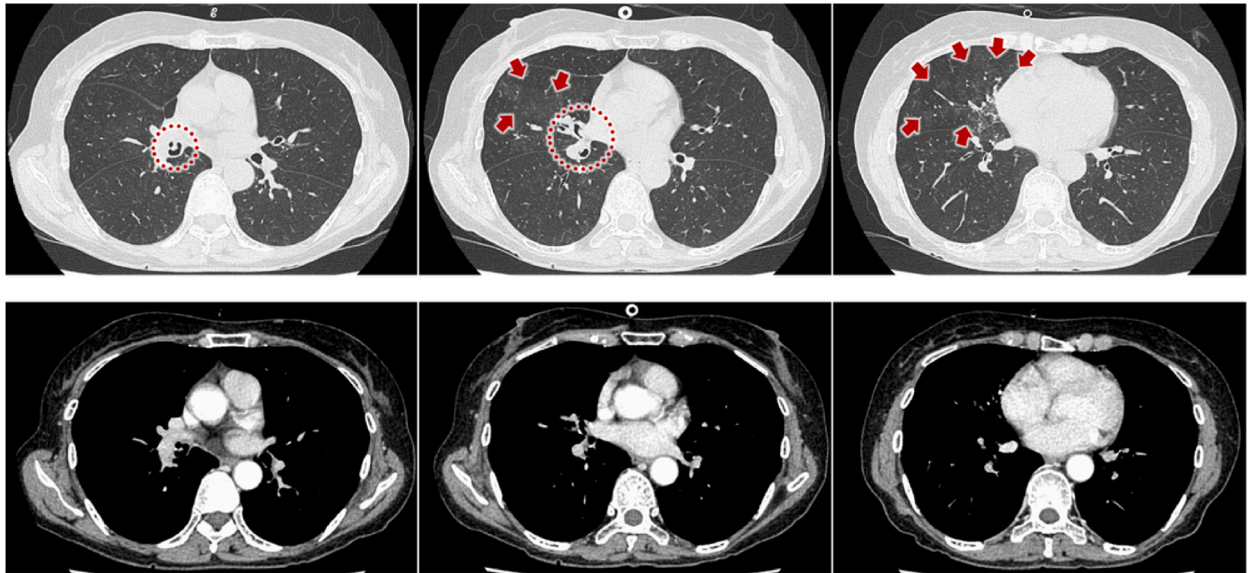


Fig. 1. Contrast-enhanced CT showed GGA in the right middle lobe (arrows) and endobronchial shadow in the right upper bronchial entrance (dotted area). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

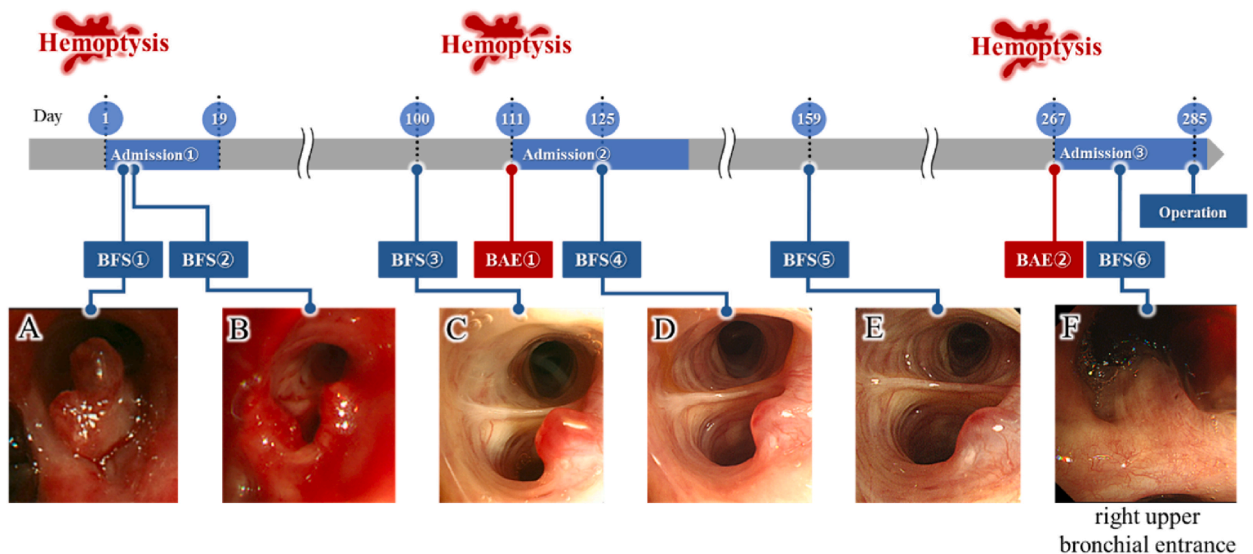


Fig. 2. Bronchofiberscopy (BFS) revealed an elevated lesion that resembled a malignant tumor (A). BFS after a biopsy shows that the lesion shrank (B). Three months after hospitalization, the patient required intensive care, and BFS revealed that the lesion had increased in size (C). BFS after BAE showed that the lesion had shrunk; however, a residual lesion was still present (D-E). Endobronchial lesions could not be observed because of the large amount of blood clots (F). BAE, bronchial arterial embolization, BFS, bronchofiberscopy. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

ter intubation showed that the lesion had shrunk (Fig. 2B). Moreover, the pathological specimen showed no evidence of malignancy. The patient was discharged on day 19 of admission without hemoptysis recurrence.

Supplementary video related to this article can be found at <https://doi.org/10.1016/j.rmcr.2022.101762>

2.1. Movie 1

Massive bleeding appeared after a biopsy of the lesion with forceps.

Three months after discharge, bronchoscopy revealed that the lesion had increased in size (Fig. 2C). Although BAE was considered an additional treatment for preventing hemoptysis, massive hemoptysis recurred two weeks after bronchoscopy, and the patient was admitted to the emergency department again. Angiography of the right bronchial arteries indicated enlarged and convoluted bronchial arteries with pulmonary artery shunts (Fig. 3A). Based on angiography findings and the absence of pulmonary diseases, she was diagnosed with primary RHBA, and BAE using a gelatin sponge was performed (Fig. 3B). After BAE, her symptoms immediately improved. Bronchoscopy after BAE showed that the lesion had shrunk; however, a residual lesion was still present (Fig. 2D and E).

Although hemoptysis had not recurred for approximately 5 months after BAE, she was admitted to our hospital again for the third time because of the recurrence of hemoptysis. Selective angiography of the right bronchial arteries revealed that the right bronchial arteries were enlarged and convoluted, and shunts were re-established between the bronchial and pulmonary arteries. In addition, the contrast medium leaked out of the lesion in the bronchus, and it was discharged by coughing (Fig. 3C, D, 3E; Movie 2). BAE using gelatin sponges was performed again on the right bronchial arteries. Although bronchoscopy after the second BAE revealed no active bleeding, the endobronchial lesion could not be observed because of massive blood clots (Fig. 2F). Because the patient wanted a curative treatment, a thoracoscopic right upper lobectomy was performed to prevent recurrence of hemoptysis. There were no complications during or after the operation. On gross appearance, endobronchial lesions were not observed in the surgical specimens (Fig. 4A and B). Although the peribronchial arteries were embolized with embolic material (Fig. 4C), red blood cells accumulated in the alveoli (Fig. 4D). This suggests that some bleeding may have persisted. In addition, minute meningothelial-like nodules were incidentally found in the peripheral lung (Fig. 4E and F). Eleven months after the operation, hemoptysis has not recurred. In reporting this case, informed consent was obtained from the patient.

2.2. Movie 2

Angiography of the right bronchial arteries indicated enlarged and convoluted bronchial arteries with pulmonary artery shunts. In addition, the contrast medium leaked out of the lesion in the bronchus, and it was discharged by coughing.

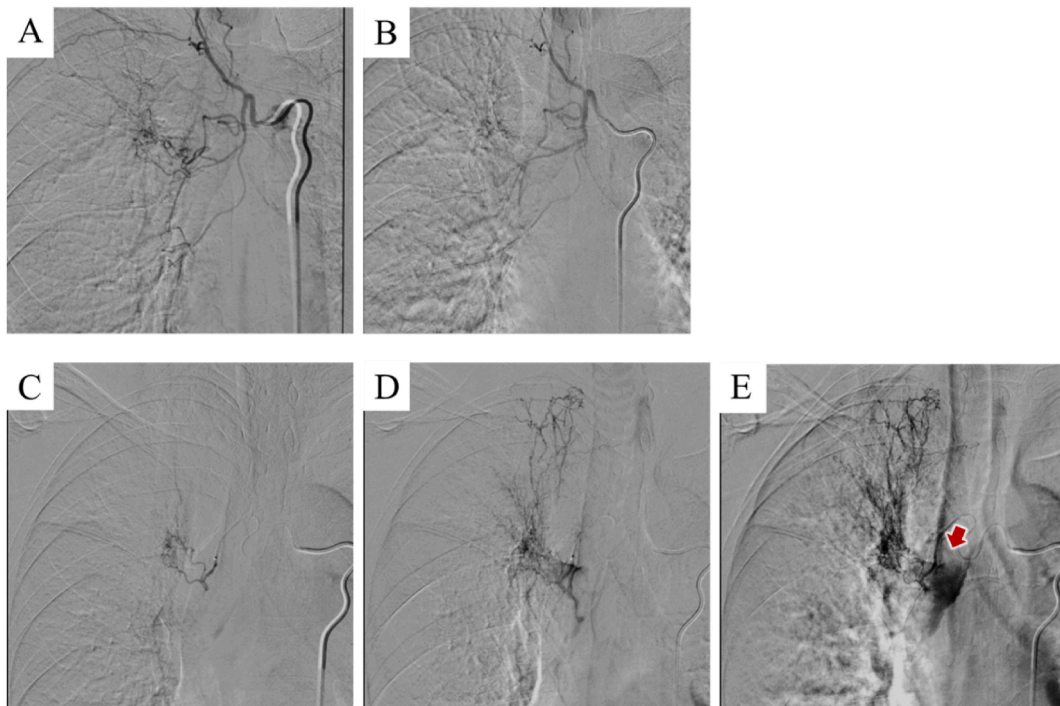


Fig. 3. Angiography of the right bronchial arteries showed enlarged and convoluted bronchial arteries with pulmonary artery shunts (A). After the first BAE, abnormal vessels disappeared (B). Five months after the first BAE, angiography of right bronchial arteries revealed that the right bronchial arteries had developed and had shunts between the bronchial and pulmonary arteries again. In addition, the contrast medium leaked out of the lesion in the bronchus and was discharged by coughing (arrow) (C-E). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

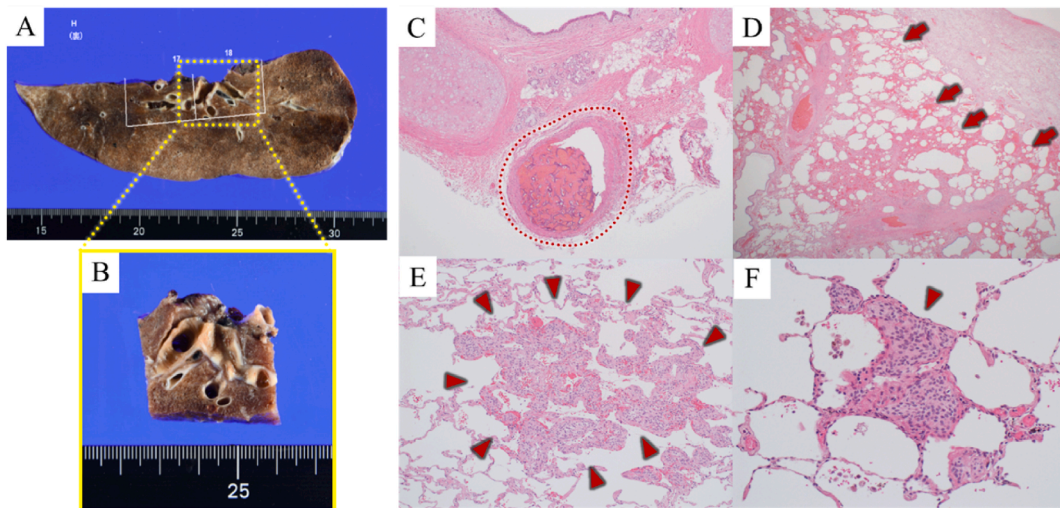


Fig. 4. On gross appearance, endobronchial lesions were not observed in the surgical specimens (A-B). The peribronchovascular arteries were embolized with embolic material (dotted area) (C). Accumulation of red blood cells was observed in the alveoli (arrows) (D). Minute meningothelial-like nodules were found in the peripheral lung (arrowheads) (E-F). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

3. Discussion

We present a case of RHBA with endobronchial lesions that had recurrent hemoptysis and was initially treated with BAE. However, hemoptysis kept recurring, and thoracoscopic right upper lobectomy was finally performed to prevent recurrence.

RHBA may show endobronchial lesions that have shapes similar to that of tumors (Fig. 2A) and can cause massive bleeding on biopsy. Bronchoscopic findings of RHBA have been previously reported, in which polypoid or other elevated lesions covered by normal mucosa are smooth-surfaced, tense, hemispherical, and sometimes pulsatile [2]. The findings of this case were similar to the characteristics of Dieulafoy's disease. Dieulafoy's disease mainly occurs in the gastrointestinal tract and is extremely rare in the bronchial tree [6], although there is no established definition of Dieulafoy's disease at present. RHBA and Dieulafoy's disease may have similar pathologies; however, the similarities or differences between these two diseases require further investigation. In this case, a biopsy was performed because the endobronchial lesion resembled a tumor, and massive bleeding occurred after the biopsy was performed. Biopsies of vascular lesions, including RHBA, should generally be avoided because it can result in massive bleeding. Although compression using forceps and ultrasound endoscopy can be effective in differentiating vascular lesions [7], the endobronchial lesions of RHBA can cause massive bleeding even with the use of a saline solution spray [3]. Contrast-enhanced CT or angiography should be considered if there is the slightest suspicion of RHBA.

Additional treatment should be considered when endobronchial lesions remain because of the possibility of recurrent hemoptysis. Although the short-term success rate of BAE for vascular lesions causing hemoptysis is about 80–90% [5], long-term outcomes have not yet been evaluated. The endobronchial lesion did not disappear, and increased in size despite the first BAE in this case. Hemoptysis stopped after the second BAE, and the endobronchial lesion disappeared according to pathological findings. Although only the second BAE improved the bleeding, surgery was reasonable because of blood retention in the alveoli of the specimen, and other bleeding lesions may have been present. In cases where the endobronchial lesion did not disappear after BAE, additional treatment, including surgery, should be considered because of the risk of fatal bleeding.

We reviewed the literature of similar cases where BAE was performed for RHBA since 2000, and extracted the following information: age, sex, chief complaint, location of bleeding point, endoscopic findings, and treatment to control bleeding [3,5,7,9–18]. As a result, there were 16 cases of RHBA in 14 reports including the case presented in this report (Table 1). Although 13 of 16 cases (81.3%) were improved only by BAE, 3 cases (18.8%) required surgery to control bleeding in addition to BAE. There were no clear differences between the cases that required surgery and the successful cases where adequate hemostasis was achieved with BAE alone. Though bronchoscopy was performed after BAE in our case, it was not performed after BAE in the 2 previously reported cases that required surgery; therefore, it was not clear whether a residual endotracheal lesion of RHBA remained after BAE in the 2 previously reported cases. The endobronchial lesions in RHBA have been reported to disappear after a successful BAE [5], and thus residual endobronchial lesions may warrant the consideration of additional treatment.

In this case, pathological findings revealed minute meningothelial-like nodules similar to meningiomas, which are often found incidentally during lung surgery or autopsy. There have been no reports that the nodules themselves cause symptoms, and the treatment is not required. However, it should be noted that cases of malignancy complications have been reported [8]. Furthermore, in this case, there were small granular shadows in the peripheral lung when CT was reviewed retrospectively. There was no association with airway or inflammatory changes, suggesting a low relationship with RHBA. In addition, there have been no reports of complications associated with RHBA and meningothelial-like nodules; therefore, they might have been discovered incidentally. However, the relationship between RHBA and meningothelial-like nodules requires further investigation.

Table 1
Previously reported cases where BAE was performed for RHBA since 2000.

No.	Authors	Year	Age	Sex	Chief complaint	Location of bleeding point	Endoscopic findings	Treatment to control bleeding
1	Iwasaki et al.	2001	44	F	Hemoptysis	Right lower lobe	Blood clots	BAE + lobectomy
2	Mitsushima et al.	2001	42	M	Hemoptysis	Left lower lobe	Bleeding	BAE + lobectomy
3	Ishimoto et al.	2003	59	M	Hemoptysis	Left lingular bronchus	Polypoid	BAE
4	Ishimoto et al.	2003	24	F	Hemoptysis	Right B9	Polypoid	BAE
5	Narato et al.	2006	56	F	Hemoptysis	Right middle lobe	Polypoid	BAE
6	Yasumoto et al.	2007	69	M	None	Right upper lobe	Polypoid	BAE
7	Okamoto et al.	2008	22	F	Hemoptysis	Right lower lobe	Bleeding	BAE
8	Ishiguro et al.	2009	55	M	Cough	Right upper lobe	Not done	BAE
9	Sanno et al.	2009	54	M	Hemoptysis	Right B8	WNL	BAE
10	Yoshida et al.	2014	78	M	Hemoptysis	Right B3	Polypoid	BAE
11	Fujimoto et al.	2017	48	M	None	Right lower lobe	Not done	BAE
12	Takano et al.	2020	78	M	None	Left basal bronchus	WNL	BAE
13	Hashiba et al.	2021	89	F	Chest pain	Right lower lobe	Not done	BAE
14	Hashiba et al.	2021	50	M	Hemoptysis	Right upper lobe	Polypoid	BAE
15	Kawabe et al.	2021	27	M	Hemoptysis	Right middle and lower lobes	Polypoid	BAE
Our Case			72	F	Hemoptysis	Right B3	Polypoid	BAE + lobectomy

BAE: bronchial arterial embolization; F: female; M: male; RHBA: racemose hemangioma of the bronchial artery; WNL within normal limit.

In conclusion, bronchoscopic findings should be assessed well, and contrast-enhanced CT or angiography should be considered if RHBA is suspected. Additional treatment should be considered for RHBA if there is recurrent hemoptysis and residual endobronchial lesions after BAE.

3.1. Learning points

- RHBA can present with endobronchial lesions that have shapes similar to that of tumors.
- The endobronchial lesions of RHBA can cause massive bleeding. Therefore, contrast-enhanced CT and/or angiography should be considered before biopsy in case similar findings are observed by bronchoscopy.
- In case endobronchial lesions remain after BAE, additional treatment, including surgery, should be considered for RHBA because of the possibility of re-hemoptysis.
- RHBA and minute pulmonary meningothelial-like nodules may coexist. Because, the relationship between RHBA and pulmonary meningothelial-like nodules is not clear, further investigation is required.

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Declaration of competing interest

The authors declare no conflict of interest.

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