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Case Report

Intercostal-to-pulmonary arterial fistula with bronchiectasis-induced massive hemoptysis: a case report and review of the literature

Hourong Zhou[1,2*], Bin He[1*], Li Zhu[3], Rongpin Wang[2], Yu Cao[1]

[1]West China Hospital, Sichuan University, Chengdu 610041, Sichuan, China; [2]Guizhou Provincial People's Hospital, [3]Guizhou Medical University; Guiyang, China. [*]Equal contributors.

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Abstract: Systemic-to-pulmonary arterial fistulas are a rare vascular malformation. Here, the authors presented a clinical case of a 58-year-old patient diagnosed with intercostal artery-to-pulmonary artery fistula, accompanied by massive hemoptysis, and a review of the literature of clinical features with intercostal artery-to-pulmonary artery fistulas.

Keywords: Arterial fistulas, bronchiectasis, hemoptysis, angiography

Introduction

In clinical practice, systemic-to-pulmonary arterial fistulas are rare vascular malformations and may be misinterpreted as patency of the ductus arterious or arterio-venous malformation. One such type of fistula, intercostal-to-pulmonary arterial fistulas, feature an extremely low incidence rate and their clinical symptoms are atypical or absent before accidental discovery during physical examinations [1]. Massive hemoptysis results from the rupture of intercostal-to-pulmonary arterial fistula and can be life-threatening. Although not fully understood, the cause of the disease is possibly due to congenital or acquired factors such as inflammation (*Actinomycetes* or *Mycobacterium tuberculosis* infection), vegetation, trauma, or iatrogenic surgical procedures. To provide reference for early detection, diagnosis, and treatment of the disease during clinical practice, we presented a case of intercostal-to-pulmonary arterial fistula accompanied by bronchiectasis-induced massive hemoptysis. A retrospective analysis on previous literature is also provided.

Case report

The patient, a 58-year-old female, presented with hemoptysis without significant inducement for 4 days. Hemoptysis was mixed with phlegm, appeared bright red with a volume of 1,000 mL, and was accompanied by discomfort such as chest distress and acratia. No fever, chest pain, abdominal pain, or hematochezia was observed. The patient presented history of tuberculosis but no recurrence was observed after one-year of anti-tuberculosis treatment. Upon physical examination, her blood pressure was 66/34 mm Hg with a heart rate of 67 bpm. Breathing sounds of both lungs were rough and a small quantity of moist rales was heard. The heart border was not enlarged. Rhythm was regular and no pathological murmur was heard from auscultatory valve areas. The abdomen was soft without any abnormality. No edema was found in bilateral lower limbs. In assistant examination, no abnormality was detected in emergent electrocardiogram. Blood routine assessment showed high leukocyte count. Serum procalcitonin examination revealed no abnormality. During chest computed-tomography (CT), significant widening was observed in the bronchus lumen of the superior lobe of the right lung. The bronchus wall showed thickening. Solid change and an exudative lesion wered observed in peripheral areas. Multiple scattering and patchy-like fuzzy shadows were detected inside the remaining area of the right lung. Bronchiectasis combined with an infection was detected in CT diagnosis and hemoptysis was found in the right lung (**Figures 1** and **2**). Treatments included

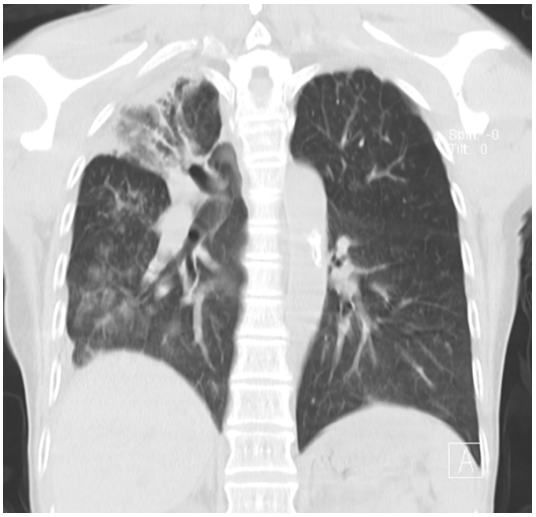

Figure 1. Reconstructed image for coronal view of CT plain scan revealed bronchiectasis in the superior lobe of right lung, solid changes and exudative lesions in peripheral areas and multiple scattering patchy-like fuzzy shadows inside remaining areas of the right lung.

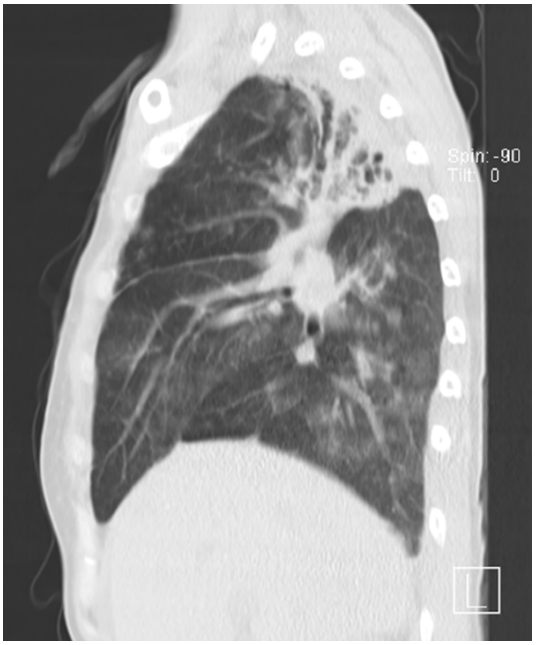

Figure 2. Reconstructed image for sagittal view of CT plain scan revealed signs similar to those detected in coronal view.

intravenous drip of levofloxacin hydrochloride (0.4 g) for anti-infection and etamsylate (3 g) for hemostasis. Continuous intravenous pumping of foliosine (1 u/h) and fluid resuscitation were implemented after hospitalization. Noradrenalin (0.5 μg/min/kg) was also administered to maintain blood pressure at 90-100/50-60 mmHg. Hemoptysis remained in remission. Descending aortic angiography under intervention was carried out and intercostal-to-pulmonary arterial fistulas in the 4th intercostal artery were observed in multiple branches of the right lung. Selective angiography was implemented again for the 4th intercostal artery. Significant thickening, circuity, branch increase, and disor-

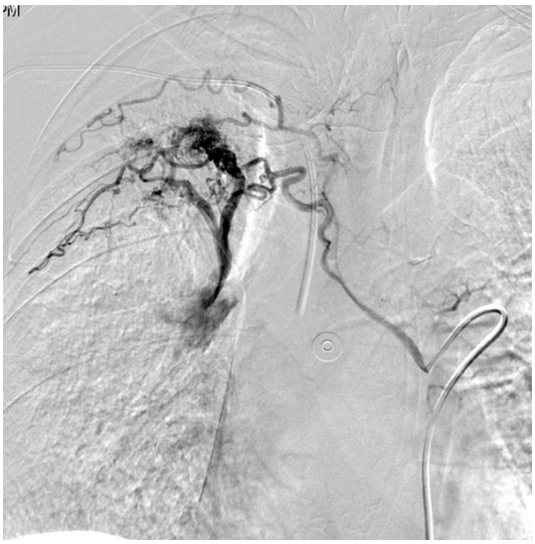

Figure 3. Selective angiography for the 4th intercostal artery revealed significant thickening, circuity, increased branching, and disorder in intercostal arteries and direct drainage of contrast agent into right upper pulmonary artery.

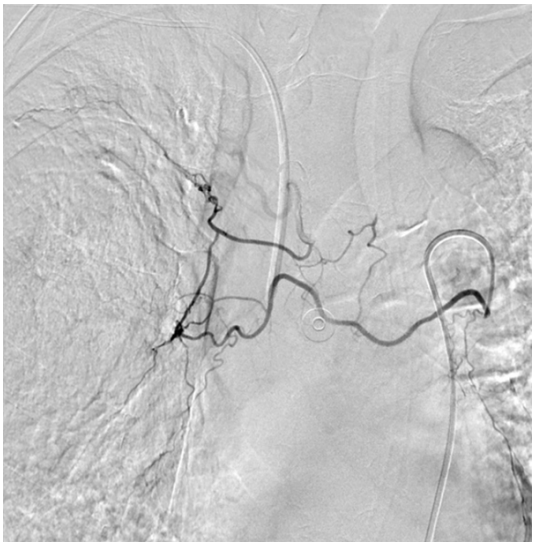

Figure 4. Selective angiography for right bronchial artery showed no abnormality in sizes of bronchial artery and its branches.

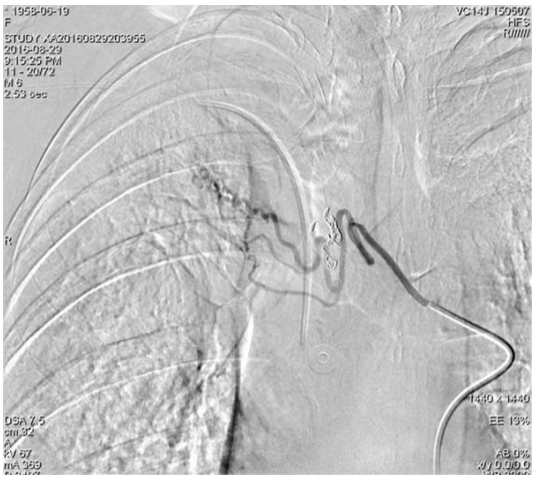

Figure 5. Abnormally thickened and disorganized bronchial arteries disappeared after embolization with detachable coils.

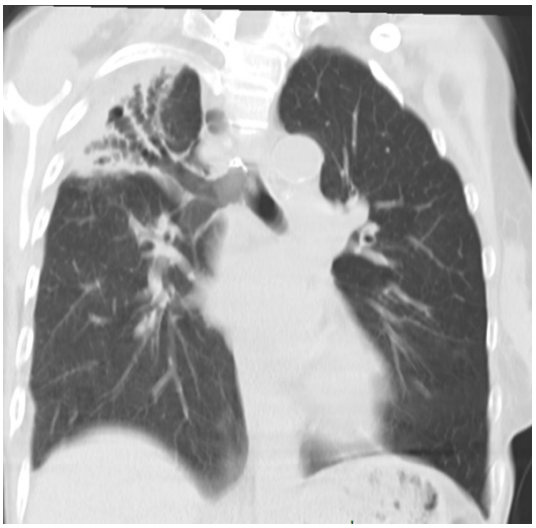

Figure 6. Reconstructed picture of coronal view of CT plain scan after treatment revealed that bronchiectasis was still present in the right upper lung, but most of the peripheral exudative lesion had been absorbed. Multiple patchy exudative lesions originally observed in lungs were absorbed.

der were observed in the blood vessels. Direct drainage of contrast agent into the right upper pulmonary artery was also observed (**Figure 3**). Bronchial arteriography revealed normal bronchial arteries on both sides and their branches (**Figure 4**). Intercostal arterial embolization under a TV monitor was implemented after diagnosis of intercostal-to-pulmonary arterial fistulas in multiple branches of the right lung, by *digital subtraction angiography (*DSA). Four pieces of MWCE-18S-3.0-4-HILAL detachable coils and four pieces of MWCE-18S-4/2-TORNADO detachable coils were used to embolize thickened and circuitous intercostal arteries. The post-operative angiography showed significant reduction in intercostal-to-pulmonary arterial fistulas of multiple branches in the right lung (**Figure 5**). CT pulmonary angiography and pulmonary plain scan were reexamined two weeks later and revealed the following results: bronchiectasis was still detectable in the upper right lung and the peripheral exudative lesion was absorbed. Multiple patchy exudative lesions originally observed in the lung were absorbed (**Figure 6**). Multiple sputum examinations showed the absence of acid-fast bacillus. The results of erythrocyte sedimentation rate and tuberculin test were negative. Anti-infection treatments lasted 2 weeks and discharge was managed after stabilization of the patient's condition. No hemoptysis was detected in follow up visits up until the present and daily activities were allowed.

Discussion

Pulmonary tissues are supplied by dual blood origins, pulmonary and bronchial arteries. The pulmonary artery system completes blood-oxygen transfer, whereas the bronchial artery system is responsible for nourishment of pulmonary supporting structures. The pressure of bronchial arteries is nearly 10 times higher than that of pulmonary arteries. In a normal state, the two vascular systems exhibit a reticular structure and feature no direct connection during communication. Systemic-to-pulmonary arterial fistulas occur in an abnormal state. Intercostal-to-pulmonary, bronchial-pulmonary, and internal mammary-to-pulmonary arterial fistulas have been commonly observed. Potential life-threatening circumstances include volume overload, pulmonary arterial hypertension, right heart failure, infectious complications, and/or massive hemorrhage caused by rupture [2]. Dyspnea, hemoptysis or heart failure is commonly observed in patients with pulmonary arterial fistula. Some patients present with no symptoms except for continuous murmuring during cardiac auscultation.

Intercostal-to-pulmonary arterial fistula cases are extremely rare. So far, we have only discovered 15 reported cases [3-17] after searching the literature database (**Table 1**). Causes of the disease include congenital abnorma-

Table 1. Case reports of intercostal-to-pulmonary arterial fistula

No.	Sex	Age (year)	Disease cause	Initial symptom	Lesion range	Fistula location	Diagnostic method	Treatment method	References
1	F	8	*Actinomycetes* infection	Back pain, acratia, emaciation, continuous murmur (grade 3/6) in cardiac auscultation.	Intercostal-to-pulmonary arterial fistula	Single side/left	Chest X-ray radiography	Drugs/embolization	Knoepfli [3]
2	M	17	Pulmonary tuberculosis	Hemoptysis	Multiple intercostal-to-pulmonary arterial fistulas	Single side/left	DSA	Pulmonary lobectomy	Syme [4]
3	F	18	Unknown	None, systolic murmur (phase 2/6) in cardiac auscultation	Intercostal-to-pulmonary arterial fistula	Single side/right	DSA	Undescribed	Kobayashi [5]
4	M	29	Unknown	None	Intercostal-to-pulmonary arterial fistula	Single side/left	DSA	Embolization	Shinichi [6]
5	M	44	Congenital cause	None, continuous murmur (grade 3/6) in cardiac auscultation	Intercostal-to-pulmonary arterial fistula	Single side/left	UCG, DSA	Undescribed	Izumi [7]
6	M	23	Trauma	Chest pain	Intercostal-to-pulmonary arterial fistula	Single side/right	DSA	Transcatheter embolization	Kubota [8]
7	M	51	Tuberculous pleuritis	Left heart failure	Intercostal/bronchial-to-pulmonary arterial fistula	Single side/left	CTA	Transcatheter embolization	Chino [9]
8	M	44	Pulmonary tuberculosis	Hemoptysis	Intercostal/internal mammary-to-pulmonary arterial fistula	Single side	DSA	Transcatheter embolization	Akahane [10]
9	M	18	Congenital cause	None, continuous mechanical murmur (grade 3/6) in cardiac auscultation.	Intercostal-to-pulmonary arterial fistula	Single side/left	CTA	Transcatheter embolization	Cantasdemir [11]
10	M	31	Unknown	None	Intercostal/internal mammary/splenic-to-pulmonary arterial fistula	Single side/left	CTA	Embolization faliure, pulmonary lobectomy and fistulectomy	Itano [12]
11	M	31	Unknown	None, abnormality was found in the chest picture obtained during physical examination.	Intercostal/internal mammary/gastric-to-pulmonary arterial fistula	Multiple sides	CTA	Multiple vascular embolization	Hideki [13]
12	M	38	Trauma	Hemoptysis	Intercostal-to-pulmonary arterial/venous fistula	Multiple sides	CTA	Transcatheter embolization	Kim [14]
13	M	35	Unknown	Dyspnea	Intercostal-to-pulmonary arterial fistula	Single side/left	Chest X-ray radiography, CTA	Undescribed	Morais [15]
14	M	39	Hereditary hemotthagic telangiectasis	None, murmur in cardiac auscultation	Dural arteriovenous fistula, Intercostal/internal mammary/inferior phrenic-to-pulmonary arterial fistula	Multiple sides	CTA	Transcatheter embolization	Miyamoto [16]
15	M	28	Unknown	None	Intercostal-to-pulmonary arterial fistula	Single side/right	CTA	Undescribed	Koc [17]

Abbreviation: F, female; M, male; DSA, Digital subtraction angiography; CTA, CT angiography; UCG, ultrasonic cardiogram.

lity: 13.33% (2/15 case-times); tuberculosis: 20.00% (3/15 case-times); trauma: 13.33% (2/15 case-times); *Actinomycetes* infection: 6.67% (1/15 case-times); and hereditary *hemorrhagic* telangiectasis: 6.67% (1/15 case-time). An unknown cause is 40.00% (6/15 case-times). *Mycobacterium tuberculosis* infection can induce intercostal-to-pulmonary arterial fistula. Inflammatory reaction in response to infections may result in increased pulmonary capillary permeability and blood cell infiltration into alveolae and occur in local lungs due to *Mycobacterium tuberculosis* toxins and massive sensitizers. Lesion tissue necrolysis caused by caseous necrosis may lead to erosion and damage of blood vessels. Bronchiectasis accompanied by twisted hemangiectasis occurs in circular vessels of the bronchus and collateral circulation increases. As a result, anastomosis between bronchus and pulmonary circulation increases. Damage, erosion, and ulceration occur in bronchus wall mucosa and hemorrhaging results from capillary damage of granulation tissue [18]. The main causes of bronchiectasis combined with hemoptysis include bronchial and non-bronchogenic artery dilatation, terminal branch anastomosis of bronchial-pulmonary artery, pulmonary arterial abnormality, and rupture of bronchial artery aneurysm. In the present case, pulmonary CT after hospitalization revealed right-side bronchiectasis (**Figures 1** and **2**). Intercostal-to-pulmonary arterial fistulas in multiple branches of the right side were identified by DSA (**Figure 3**) but the bronchial-pulmonary arteriography suggested a normal state of bronchial arteries in the right side (**Figure 4**). The intercostal artery arteriography after embolization revealed the disappearance of thickened and circuitous intercostal arteries and original pulmonary arterial fistula (**Figure 5**). A CT image of the lung after the 20-day treatment showed that bronchiectasis was at the apicoposterior segment of the superior lobe of the right lung with absorption of multiple scattering and patchy-like fuzzy shadows appeared in bilateral lungs (**Figure 6**). These results further provide evidence that hemoptysis in the patient was not caused by bronchiectasis.

Patients with intercostal-to-pulmonary arterial fistulas show no specific symptoms. Clinical manifestation partially depends on fistula orifice size, function between fistula orifice and connecting blood vessels, and distance between fistula orifice and the heart. Our literature review [3-17] showed the following symptoms: hemoptysis: 20.00% (3/15 case-times); pain: 13.33% (2/15 case-times); dyspnea: 6.67% (1/15 case-time); left heart failure: 6.67% (1/15 case-time); asymptomatic: 53.33% (8/15 case-times); and accompanying continuous cardiac murmur: 33.33% (5/15 case-times). Pathogenic sites of intercostal-to-pulmonary arterial fistulas are mostly found in a single side (80.00%, 12/15 case-times) and more commonly on the left side (60.00%, 9/15 case-times). Only in 20.00% of cases (3/15 case-times) were the fistulas observed on both sides. Sex ratio of the 15 cases was 6.5:1 (13 male and 2 female cases).

Primary diagnostic methods for intercostal-to-pulmonary arterial fistulas include chest X-ray examination, DSA, CT angiography (CTA), ultrasound, and magnetic resonance imaging. Incisura below the rib and pulmonary infiltration shadows caused by hemorrhage are observed in X-ray examinations. The clinical value of a chest X-ray examination is limited and cannot be used as the first choice. DSA, which can clearly display site, quantity, shape, and homodynamic characteristics of intercostal-to-pulmonary arterial fistulas, is the gold standard for diagnosis. Embolic hemostasis can also be carried out on the hemorrhage site simultaneously with DSA examination. However, these methods may result in high costs, multiple complications, and extensive damage. CTA is noninvasive angiography with high accuracy and features less trauma and radiation in comparison with DSA. CTA, as a routine angiography, has become the preferred method for diagnosis of pulmonary vascular diseases. Although an ultrasound examination does not cause radiation damage [18], its detection range is limited. Intrapulmonary vascular disease can be clearly displayed by magnetic resonance angiography for pulmonary arteries and the detection rate with this method can be improved without trauma. Magnetic resonance angiography may replace pulmonary artery DSA [19]. Diagnostic methods used in collected cases are: DSA, as the gold standard, used in 40.00% of the cases (6/15 case-times) and combined detection with ultrasonic cardiogram was used in 6.67% of cases (1/15 case-time). A total of 53.33% of patients (8/15 case-times) selected CTA, which features less trauma and a lower cost. Pulmonary vascular CTA is recommended as the preferred method for diagnosis of intercostal-to-pulmonary arterial fistulas.

Several treatment methods are available for intercostal-to-pulmonary arterial fistulas but, currently, there is not a unified standard. Pulmonary lobectomy and fistula ligation have been frequently used in the past. However, those methods resulted in major traumas with slow postoperative recuperation. With the continuous development of intervention techniques, local arterial embolization, presenting minimal trauma with quick and complete hemostasis and definite short- and long-term therapeutic effects, has become the preferred method in clinics [20]. Pulmonary artery embolization was used in 60.00% (9/15 case-times) of the cases collected for this study. Pulmonary lobectomy and fistula ligation, which can cause major trauma and slow recovery, were used in 13.33% (2/15 case-times) of the cases. Conservative drug treatment was used in 6.67% (1/15 case-time) of cases.

We, therefore, conclude that pulmonary vascular CTA should be carried out to clarify the diagnosis when continuous cardiac murmuring accompanied by hemoptysis is observed during a clinical visit. Bronchial and intercostal arteriography should be implemented on time when a high amount of hemoptysis is observed and when drug treatment proves to be ineffective. Embolization is required after confirmation of the hemorrhage site. Pulmonary lobectomy and fistula ligation should be performed when high pulmonary arterial pressure is revealed in postoperative monitoring and when continuous hemoptysis occurs after embolization.

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Disclosure of conflict of interest

None.

Address correspondence to: Yu Cao, West China Hospital, Sichuan University, 37 Guo Xue Xiang, Chengdu 610041, Sichuan, China. E-mail: dr.yu.cao@gmail.com

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