Diagnoses and treatment decision in anomalous systemic arterial supply to basal segments without sequestration

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Abstract: Objectives: To conclude the diagnoses and treatment decision in anomalous systemic arterial supply to basal segments without sequestration. Methods: Clinical manifestations, CT scanning, diagnoses and treatment of 6 cases from 2010 to 2013 were reported together with literature review. Results: All 6 cases were diagnosed by high resolution CT, and were confirmed in operation. It presented clinical features as male gender and hemoptysis. CT showed one anomalous systemic artery arising from descending thoracic aorta supplying the left lower lobe. 1 case who was comitant with von Willebrand disease survived after endovascular embolization, 4 of 5 cases were cured by surgical treatment and 1 died of postoperative bleeding. Conclusions: Definite diagnoses can be made by means of high resolution CT. For patients with complicated condition, endovascular embolization could be a safer alternative.

Keywords: Anomalous systemic arterial supply, high resolution computed tomography, video-assisted thoracoscopic surgery, endovascular embolization

Introduction

Anomalous systemic arterial supply to basal segments without bronchial pulmonary sequestration is a rare congenital anomaly. However, it has arisen more attention as the use of high-resolution CT (HRCT) in these years. The treatment modality is still controversial regarding whether endovascular embolization is the first choice for all cases. We reported 6 patients who were diagnosed by HRCT and treated by operation, then literature was reviewed.

Case 1

A 31-year-old male was admitted to our hospital because of repeated hemoptysis for 7 months. About 5 mL bright red blood was coughed out each time when hemoptysis occurred. There was no fever, night sweat, palpitation, chest pain, pus sputum and breathlessness. He used to suffer gingival bleeding, subcutaneous hemorrhage and epistaxis since childhood, even had upper gastrointestinal hemorrhage once. He was non-smoker, denied positive family history. His life signs were stable. There was no positive sign in physical examination.

In blood studies, no inflammatory findings were noted. Platelet test was also normal. However, activity of coagulation factors decreased. Hemophilia screening tests showed that coagulation factor VIII activity was 28% (reference range from 60% to 150%), factor IX activity was 62% (reference range from 60% to 150%), VWF activity was 18% (reference range from 50% to 160%), factor XI activity was 74% (reference range from 60% to 140%), and factor XII activity was 46% (reference range from 60% to 140%). During bronchoscopy examination, the patient continued expectorating dark blood from bronchi in left lower lobe, but his bronchial tree was normal without sequestration. Contrast-enhanced chest CT and volume reconstruction (VR) were performed, the result showed a dilated anomalous artery (15 mm in diameter) arising from lateral aspect of descending aorta supply to left basal segments (Figure 1).
Diagnoses and treatment of anomalous systemic arterial supply

The patient's final diagnoses was anomalous systemic arterial supply to left basal segments comitant with von Willebrand disease.

The patient was prescribed with fresh frozen plasma and cryoprecipitate to stop hemoptysis. He refused any surgical or endovascular therapy because of potential complications in the beginning. Then he accepted endovascular embolization with coil in Nanfang Hospital Affiliated to Southern Medical University one year later. The operation was successful. The anomalous systemic artery was completely obstructed. Hemoptysis ceased and no side effect emerged during follow-up for more than 5 years. As far as we knew, this was the first case reported of such a anomaly comitant with von Willebrand disease and cured by embolization with coil.

Case 2

A 29-year-old male presented with recurrent hemoptysis for 5 years. About 15 mL bright red blood was coughed out each time for three consecutive days. He was non-smoker, and had no special past history. Physical examination and laboratory findings showed no abnormalities. Bronchoscopy revealed no abnormality in bronchial tree.

Chest CT showed that a dilated anomalous artery arising from lateral aspect of descending aorta supplying to left basal segments, the interlobar artery distal to the origin of the superior segmental artery was absent, and venous return was via the large inferior pulmonary
vein into the left atrium (Figure 2).

The patient underwent left lower lobectomy with ligation of the anomalous artery by Video-assisted Thoracoscopic Surgery (VATS) one year later in the First Affiliated Hospital of Guangzhou Medical University. The operation was uneventful. No hemoptysis recurred during follow-up.

Case 3

A 27-year-old male, non-smoker, was referred to our hospital because of repeated cough and bloody sputum for 8 months. There was no fever, chest pain and tightness. He accepted antibiotics therapy including azithromycin and levofluxacin, but it was in vain. His past history was negative. Physical examination and laboratory findings were normal as well as bronchoscopy examination. Chest CT scanning found anomalous systemic arterial supplying to normal basal segments of the left lower lobe (Figure 3).

The patient accepted left lower lobectomy with ligation of the anomalous artery by VATS in the First Affiliated Hospital of Sun Yat-sen University. The operative course was satisfactory and he was asymptomatic in follow-up.

Case 4

An 18-year-old non-smoker male was referred to our hospital because of cough and hemoptysis for 2 months. Hemoptysis ceased after haemostasis and anti-infection therapy in local hospital. He had no tuberculosis and congenital heart disease. Physical ex-
amination and laboratory findings were within normal range at admission. Bronchoscopy showed no bronchial pulmonary sequestration. Chest CT showed a sigmoid-shaped anomalous systemic artery originating from the lower descending thoracic aorta with three branches supplying to basal segments of the left lower lobe (Figure 4).

Left lower lobectomy with ligation of the anomalous artery was performed under VATS in our hospital. The patient recovered successfully. Hemoptysis disappeared during follow-up.

Case 5
A 23-year-old male, non-smoker, was presented to our hospital with complain of cough, bloody sputum and high fever for one month. He coughed out about 30-40 mL bright red blood each time for 3 days before hospitalization. The patient had no special past history. He had normal physical examination and laboratory findings at admission. Colour Doppler ultrasound showed an arterial branch from descending aorta. Bronchoscopy examination was normal. Anomalous systemic arterial supply to basal segments in the left lower lobe without sequestration was demonstrated by Chest CT (Figure 5).

No surgical contraindication was found and the patient underwent left lower lobectomy with ligation of the anomalous artery by VATS in our hospital. Regrettfully, the patient died of complication of postoperative bleeding.

Case 6
A 54-year-old non-smoker female was presented to our hospital because of repeated hemoptysis for 4 years. Hemoptysis could be ceased without treatment each time. Her past history was negative. There was no positive sign in physical examination. Bronchoscopy showed no abnormal findings. Chest X-ray showed shadow in left lower lobe. Chest CT showed anomalous systemic artery supplying to left basal segments of the lower lobe (Figure 6).

Left lower lobectomy with ligation of the anomalous artery was performed under VATS in Guangdong General Hospital. The patient’s postoperative recovery was uneventful, and she was free from hemoptysis during follow-up.

Results
Case summaries
The clinical features of the 6 patients are summarized in Table 1. SPSS 20 software was utilized for the inputting, tabulation, and statistical analysis.

Clinical data
The median age of the 6 patients were 25 years (range 18-54). Five cases (83.3%) were young male and one (16.7%) was old female. All of them complained hemoptysis and 4 patients (66.7%) complained persistent cough simultaneously. Patient 1 had a comitant disease of von Willebrand disease diagnosed this time. Physical examinations were normal. Cardiac murmurs or abnormal electrocardiogram findings were not detected in any of the patients. Decrease of coagulation activity was found in patient 1, there were no special findings in laboratory test in other patients. Bronchoscopy in all 6 cases showed no bronchial sequestration.

All of 6 patients’ final diagnoses were made as anomalous systemic arterial supply to the basal segments without pulmonary sequestration by means of medical imageology, using 320-detector CT with volumen reconstruction in our hospital, and were confirmed by surgery or angiography. In all cases, imageology showed that the left basal segments were supplied by one anomalous artery, which arose from the descending thoracic aorta, without pulmonary arteries to the basal segments. The anomalous artery was 6-15 mm in diameter (with a median of 10 mm). No direct communications between the anomalous systemic artery and the veins of the basal segments were noted, and then the blood drained into the left atrium via the large inferior pulmonary vein.

Left lower lobectomy with ligation of the anomalous artery by VATS was performed in patients 2-6. Four cases showed a good prognosis. Regrettfully, patient 5 died of postoperative bleeding. Patient 1 who had comitant disease of von Willebrand disease underwent endovascular embolization with coil 1 year later and achieved a good result.

According to the data of patients 4 and 5, marked capillary dilatation was seen on the vis-
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### Table 1. Clinical features of the patients

<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
<th>Case 5</th>
<th>Case 6</th>
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DTA, descending thoracic aorta; LIPV, left inferior pulmonary vein; NA, not acquired; vWD, von Willebrand disease.
ceral pleural membrane of the basal segment during operations, without pleural adhesion. Histopathological examination showed that the anomalous artery was elastic vessel, tissue around the artery was rich in collagen fiber. Pulmonary consolidation caused by edema, partial congestion and bleeding was seen. Hemodiderin-laden-macrophages and lymphocytes were noted in the alveoli and interalveolar tissue without fibrous hypertrophy. Alveolar hemorrhage in histological pathology was in accordance with hemoptysis clinically.

Discussion

Anomalous systemic arterial supply to the basal segments without sequestration was a very rare congenital anomaly. It was regarded as Pryce type I sequestration [1]. However, this anomaly has normal bronchial connections and the involved lung is not sequestrated. More and more data showed that anomalous arterial supply was different congenital anomaly from true sequestration [2]. With the rapid development of HRCT and magnetic resonance imaging (MRI), the number of diagnosed and reported cases is increasing. Consensus of diagnoses and treatment is more important than the category of this anomaly, that’s why we reviewed this literature.

WU discovered such anomaly was related to Asian ethnicity [3]. Most cases reported were Japanese, Chinese and Indian. Hemoptysis, cough, bloody sputum and dyspnea are the most frequent clinical symptoms in adults. 100% cases we reported had hemoptysis. While the main manifestations in children are heart murmur, respiratory distress, cardiomegaly, or heart failure [4]. The diagnoses should be suspected when a patient suffered recurrent hemoptysis and cough, as well as in the presence of a continuous murmur, cardiomegaly and persistent shadow on chest radiographs, especially for Asian patients [3]. The most difficult differential diagnoses is pulmonary sequestration. Bronchoscopy examination is essential for confirming the presence or absence of a normal tracheobronchial connection.

Conventional angiography has been superseded by HRCT for diagnoses of anomalous systemic arterial supply. Chest X ray may show a vessel-like opacity. Contrast-enhanced CT scan can be diagnostic. HRCT combined with multiplanar reconstruction, maximum intensity projection and volume reconstruction consistently gain combinations of characteristic findings and perspective [5]. Just like 6 cases reported in present study, orientation and distribution of the anomalous systemic artery were precisely depicted. Most of the time, in anomalous arterial supply without sequestration, one anomalous artery arise from descending thoracic aorta, and feeding the basal segments of the left lower lobe, the venous drainage to the left atrium is via pulmonary veins, and pulmonary artery is absent [2]. Anomalous arterial supply to the right lower lobe other than the left side have been reported [6], so as the left upper lobe [7] and two aberrant arteries [8, 9]. Therefore, although there is usually only a single aberrant artery supply in left, the possibility of another aberrant artery or in other position should be considered.

Treatment is recommended for all patients suffered this anomaly with or without symptoms. Because it has potential risk of hemoptysis and heart failure. Treatment aims to relieve the volume overload on the left ventricle by abolishing the arteriovenous shunting. This can be achieved by lobectomy of the affected lung lobe, ligation of the aberrant artery and therapeutic embolization [10, 11]. Surgical procedures or embolism position would be very carefully selected.

In reviewing the literature, transarterial embolization has recently been reported more and more in anomalous systemic arterial supply. We found no complication in these reports [12]. Because of less complication and shortened recovery time, the embolotherapy is considered as a minimally invasive, safe, valuable, and more effective treatment compared with surgical intervention [13, 14]. One case reported that a cecum of the stump, which was complication of lobectomy, was repaired by endovascular stent-graft implantation [15]. Although
whether embolization is the first choice for all anomalous systemic supply is still arguable [16], it suggested that embolization was especially suitable for children [17], patients with complicated condition, which including congenital heart disease such as atrial septal defect and scimitar syndrome [18], dual abnormal arteries supply [17], aneurysm [15] and hematologic disease like our case 1.

At last, the limited number of cases in this study were not enough to fully explore the diagnoses and treatment of anomalous systemic arterial supply to basal segments. It is our duty to report and accumulate such cases.

Conclusions

Definite diagnoses can be made by means of high resolution CT, which also plays an important role in the treatment decision. For patients with complicated condition, therapeutic embolization could be a safer alternative.

Disclosure of conflict of interest

None.

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