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Olgu Sunumu / Case Report

Congenital Bronchial Artery - Pulmonary Artery Fistula in a Young Adult

Genç Erişkin Bir Olguda Konjenital Bronşiyal Arter - Pulmoner Arter Fistülü

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ABSTRACT

Primary vascular malformations such as connection between bronchial artery and pulmonary artery or pulmonary vein are quite rare. A 18-year-old male patient with the first episode of massive hemoptysis was found to have bronchial artery-pulmonary artery fistula, a rare vascular anomaly, on selective bronchial angiography. Since medical history revealed no acquired cause, it is considered congenital. He underwent bronchial artery embolization and hemoptysis was completely resolved after endovascular treatment. The presence of such vascular anomalies should be suspected in patients with massive hemoptysis. Bronchial artery embolization is a safe and effective nonsurgical treatment option for such patients.

Key words: Hemoptysis, vascular malformation, bronchial artery-pulmonary artery fistula, bronchial artery embolization.

ÖZET

Bronşial arter ve pulmoner arter veya pulmoner ven arasındaki bağlantı gibi primer vasküler malformasyonlar olukça nadirdir. İlk kez olan masif hemoptizi ile başvuran 18 yaşındaki bir erkek hastada selektif bronşial anjiografide nadir bir vasküler anomali olan bronşial arter-pulmoner arter fistülü saptandı. Öyküde hiç bir kazanılmış neden bulunmadığı için konjenital olarak düşünüldü. Olguya bronşial arter embolizasyonu uygulandı ve hemoptizisi endovasküler tedaviden sonra tamamen düzeldi. Masif hemoptizili hastalarda böyle vasküler anomalilerden şüphelenilmelidir. Bronşial arter embolizasyonu bu hastalarda güvenli ve etkili cerrahi olmayan bir tedavi yöntemidir.

Anahtar kelimeler: Hemoptizi, vasküler malformasyon, bronşial arter-pulmoner arter fistülü broşial arter embolizasyonu.

INTRODUCTION

Under normal anatomical and physiological conditions, bronchial arteries, which provides alimentary systemic circulation of the lungs, arise from aorta, proceed along with bronchial tree, and flows into pulmonary capillaries at the level of respiratory bronchioles; thus, functional pulmonary circulation is anastomosed to systemic bronchial circulation[1]. Abnormal connections can be seen between arterial and venous vascular structures of the lungs[2]. The most common pulmonary vascular anomalies include pulmonary arteriovenous fistulas, pulmonary arteriovenous malformations, pulmonary arteriovenous aneurisms, pulmonary sequestrations, and hamartoma. These lesions are usually congenital and may rarely be acquired[3]. Connections between systemic arterial circulation and pulmonary artery are called as bronchial arteriovenous malformations, bronchopulmonary arterial anastomosis, bronchopulmonary shunt, or

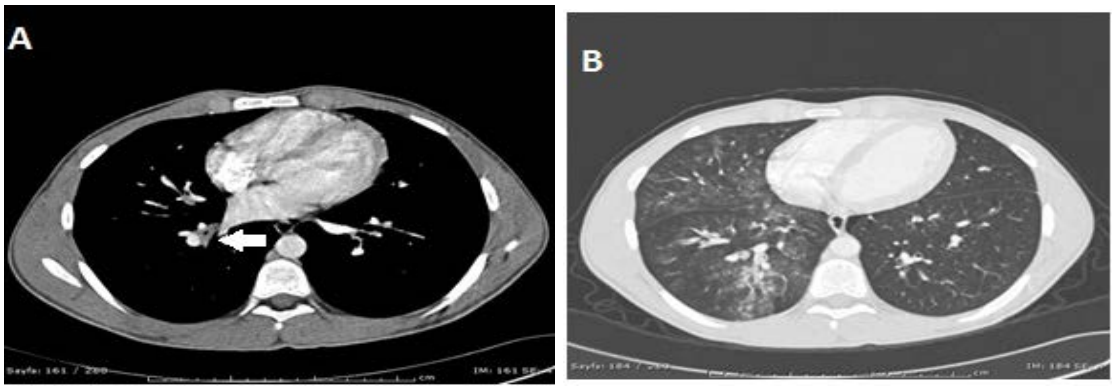
bronchopulmonary fistula. Such abnormal vascular connections can quite rarely be demonstrated radiologically, particularly in the absence of an acquired cause such as infection, trauma, surgery or malignancy[4]. They are usually asymptomatic, but rarely present with complications such as hemoptysis[5,6] Chronic inflammatory diseases of the lungs, such as tuberculosis[7,8] and pneumonia[8,9], have been defined in the literature as the common causes of connections between pulmonary and bronchial arteries.

We report a case of massive hemoptysis in a young adult secondary to a bronchial artery-pulmonary artery fistula, in the absence of an acquired cause, and complete improvement was provided by successful bronchial artery embolization.

CASE REPORT

An 18-year old male patient admitted to the emergency room with approximately 600 ml hemoptysis. Past medical and family histories of this non-smoker patient were unremarkable. General status of the patient was good, and he was cooperated. His blood pressure was 120/80 mmHg, respiratory rate was 22/min, body temperature was 36 ^{0}C, and oxygen saturation was 94%. Physical examination revealed rales in the right lower lung on auscultation. Sputum smear and culture for *Mycobacterium tuberculosis* were negative. On his laboratory analyses, results of complete blood count, blood biochemistry and urine analysis were in normal ranges, and he had no bleeding diathesis. Erythrocyte sedimentation rate was 26 mm/hour, C-reactive protein was 5.2 mg/L.

Chest X-ray revealed heterogeneous opacity in the right paracardiac area. On computed tomography (CT) of the thorax, alveolar consolidation and peribronchial thickening were detected in the right lower lobe (Figure 1A-B). Since hemoptysis persisted, bronchial angiography was planned before bronchoscopy both for diagnosis and treatment if needed. The right main femoral artery was accessed via Seldinger needle, and 6F vascular sheath was placed. The right and left main pulmonary arteries were selectively accessed and angiographic images were obtained. There was no pathology related to the left bronchial artery. However, a fistula was detected between the bronchial artery and pulmonary artery of the right lower lobe The bronchial artery ends in a network of abnormal arteries and then start filling the pulmonary artery. The network forms the fistulous component. Thereafter, distal aspect of bronchial artery was accessed via 3F microcatheter and the fistula was closed by injecting 0.2 ml of 1/6 cyanoacrilate and lipiodol mixture (Figure 1C-D). Control angiography revealed that the fistula has been completely closed (Figure E). Bronchoscopy was not performed since hemoptysis disappeared after endovascular treatment.



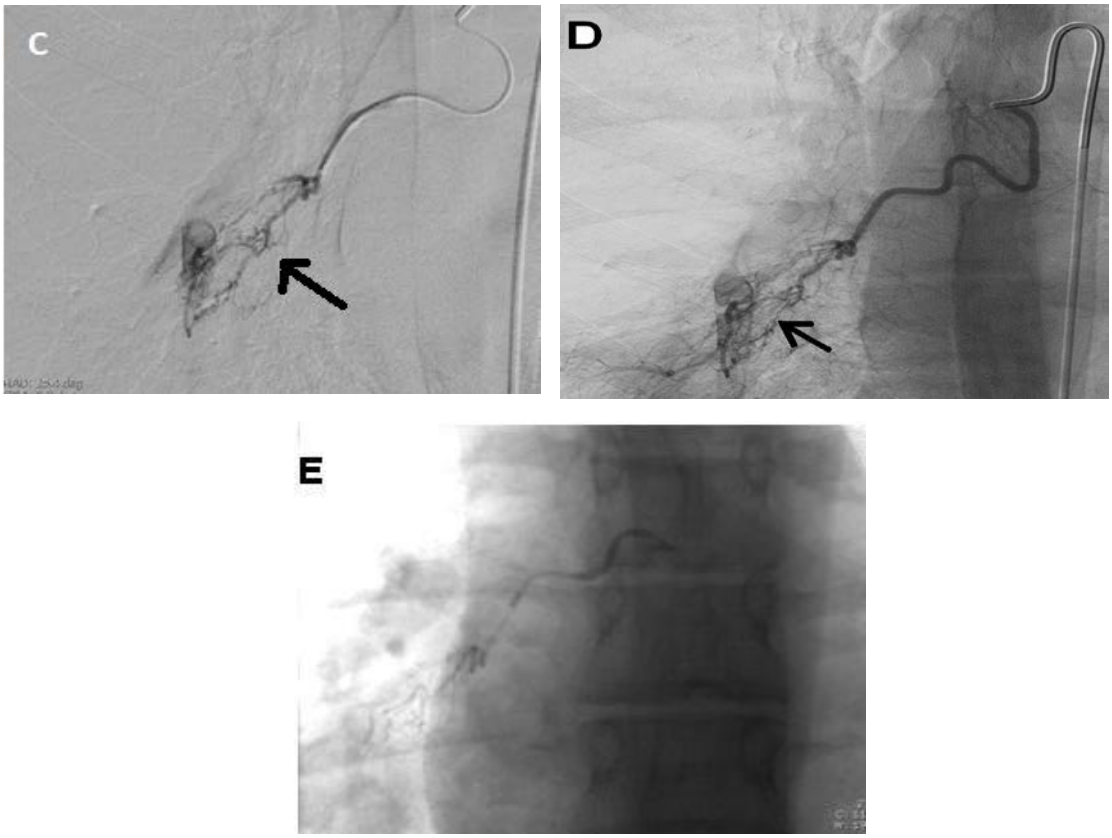


Figure1: A: CT angiography of the thorax: Peribronchial thickening in the lower zone of the right lung.
B: Ground glass opacity in the lower lobe of the right lung suggesting intraalveolar hemorrhage.
C and D: Selective and superselective bronchial arteriography: The bronchial artery ends in a network of abnormal arteries and then start filling the pulmonary artery. The network forms the fistulous component.
E: The glue cast formed after endovascular treatment with the injection of cyanoacrylate+lipiodol mixture through a microcatheter from the right bronchial artery

DISCUSSION

Blood supply of the lungs is provided by two sources. Thin-walled pulmonary arteries deliver non-oxygenated blood via low-pressure circulation into the lungs for gas exchange and oxygenated blood returns to the heart through pulmonary veins. At the same time, bronchial arteries provide nutrition and oxygen support for the lungs and bronchi via systemic pressure[10]. Classically, bronchial arteries arise from the descending aorta at the level of 5th and 6th thoracic vertebras. The lungs can receive systemic blood also via intercostal, internal mammary, esophageal and coronary arteries[11].

Bronchial arteries extend along the bronchi, join to the small pulmonary arterioles in an anastomotic plexus at the level of preterminal bronchiole and give bronchopulmonary arterioles[9,10]. Thus, normal physiological connection occurs between systemic and pulmonary vascular systems; this is the right-to-left shunt, which has no clinical importance. Under normal conditions, this arteriolar plexus is functionally closed without blood flow; these channels may be opened in case of decreased blood flow like pulmonary embolism or may develop neovascularization[10,11]. Despite the hypothesis that these vascular abnormal connections might occur as a response to an

intrauterine ischemic event, the lungs and proximal pulmonary arteries were anatomically normal in the present case.

Primary malformations including the connection between bronchial artery and pulmonary artery or pulmonary vein are quite rare. Only 4% of pulmonary arteriovenous malformations include systemic arterial connections[12]. Bronchial arterial malformations are more common in males and are usually unilateral. They most frequently occur in the right lung. The prevalence of clinical hemoptysis is 10-15%[12].

Infection is the most leading acquired cause of connections between systemic and pulmonary arteries. Bacterial pneumonia[10], tuberculosis[7,8] and pulmonary actinomycosis[13] lead to connections usually in the form of pulmonary arteriovenous fistula; rarely, connections between systemic artery and pulmonary artery may also occur. Preexisting vascular abnormal connections might lead to systemic connections with the infection of the lung area[13] Chronic inflammation of this area may cause neovascularization of the capillaries, in addition to the dilatation of normal anastomotic connections[8].

Although arteriovenous or arterioarterial fistula due to blunt trauma has been defined penetrating injuries such as stab wound or shot are the most common causes[13,14]. The fistula between internal mammary artery and pulmonary artery after coronary bypass surgery is the most frequently encountered iatrogenic fistula[15]. Concurrency of systemic and pulmonary arterial anastomoses have been defined also in the cancers including lung cancer and lymphoma[16].

An acquired cause has been determined in the majority of the cases. We considered the situation of the present case as congenital, since his history revealed no acquired cause, such as infection, trauma, surgery or malignancy.

Bronchiectasis, tuberculosis, cystic fibrosis, aspergillus pneumonia, lung abscess and heart valve diseases are the leading causes of hemoptysis particularly in young ages, whereas lung cancer and tuberculosis are the leading causes in advanced ages. Hemoptysis due to abnormal vascular connections is quite rare. It results from the connection between small airways and arteriovenous fistula[17].

Cremaschi et al.[18] discussed different etiologies in their study on bronchial artery embolization, which is one of the treatment options for hemoptysis. Bronchiectasis and tuberculosis are the most common causes in that series. Bronchial embolization has been performed for the treatment of 209 patients with massive hemoptysis; 98% have been controlled within the first 24 hours and remaining 2% have been controlled in 48 hours and the researchers reported no complication. In various series, success rates of 92% can be achieved and procedure-related complications have been reported to be quite rare[19,20].

Mechanical coils and liquid cyanoacrylate both can be used for sealing of arteriovenous fistulas. In this case there was a network of arterial vessels between the bronchial artery and the pulmonary artery. In such cases, coil embolization will block the fistulous connection in one or two points because we do not have tiny coils to embolize the whole vasculature. On the other hand, a liquid embolizing agent will diffuse through all the vasculature and block evert part of it. This in turn will help prevent new collateral formation through the patent abnormal vessels. Use of cyanoacrylate needs more experience for which our interventional radiology physicians have enough experience.

In conclusion; bronchial artery-pulmonary artery fistula is a rare vascular anomaly, which may present as massive hemoptysis. Bronchial artery angiography and embolization has become a mainstay in the treatment of hemoptysis. This procedure might be beneficial in the presence of vascular anomalies presented with massive hemoptysis, in the hands of an experienced interventionalist with appropriate digital technology. The procedure serves as both first line therapy and

as a bridge to the targeted therapies for underlying diseases. Moreover, it should be kept in mind that improvement is possible without need for invasive intervention.

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