Sistemik Dolaşımdan Beslenen Pulmoner Arteriyovenöz Fistül İle Birlikte Olan Rendu Osler Weber Sendromu

RENDU OSLER WEBER DISEASE ASSOCIATED WITH SYSTEMIC SUPPLY TO PULMONARY ARTERIOVENOUS FISTULA

Alpay Sarper, *Necdet Öz, Abid Demircan, **Erol Işın

Akdeniz Üniversitesi Tıp Fakültesi, Göğüs Cerrahisi Ana Bilim Dalı, Antalya
**SSK Antalya Hastanesi, Göğüs Cerrahisi Kliniği, Antalya*
***Antalya Göğüs Cerrahisi Merkezi, Göğüs Cerrahisi Kliniği, Antalya*

Özet

Hemoptizi, epistaksis ve çabuk yorulma şikayetleri ile hastanemize başvuran 20 yaşındaki erkek hastanın çekilen direkt toraks grafisinde düzgün kenarlı lezyon izlenmekteydi. Fizik muayenesinde oral ve nazal mukozasında multiple telanjiektaziler izlendi. Toraks bilgisayarlı tomografisinde ve pulmoner anjiyografisinde sol alt lobda arteriyovenöz fistül görüldü. Bu bulgularla hastaya Rendu Osler Weber Sendromu tanısı konuldu. Sistemik pulmoner arter ile de ilişkisi olan geniş pulmoner arteriyovenöz anevrizma lobektomi ile başarılı olarak tedavi edildi.

Anahtar kelimeler: Rendu Osler Weber sendromu, pulmoner anevrizma, herediter hemorajik telanjiektazi, arteriyovenöz fistül

Türk Göğüs Kalp Damar Cer Derg 2004;121:61-63

Summary

A 20-year-old man presented with hemoptysis, epistaxis and severe fatique. Chest X-ray film revealed an abnormal shadow. On the physical examination, multiple telangiectases were found at the oral and nasal mucous membrane and tongue. Computed tomography and pulmonary angiography showed an arteriovenous fistula of the left lower lobe. The diagnosis was Rendu Osler Weber Disease. Lobectomy was performed because of the systemic supply to the large pulmonary arteriovenous aneurysm.

Keywords: Rendu Osler Weber disease, pulmonary aneurysm, hereditary hemorrhagic telangiectasia, arteriovenous fitula

Turkish J Thorac Cardiovasc Surg 2004;12:61-63

Introduction

Connections of the pulmonary artery to the pulmonary vein resulting in aneurysmatic dilatation are defined as pulmonary arteriovenous aneurysm (PAVA) Rendu Osler Weber Disease (ROWD) or hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant disorder characterised by telangiectases and arteriovenous malformations of skin, mucous membrane, and potential each organ. Churton reported the first case of PAVA in 1897, and ROWD was set forth in 1901 by Osler [1]. We reported a rare case of ROWD associated with PAVA, which received blood supply from the bronchial artery.

Case

A 20-year-old man was admitted to the hospital with hemoptysis, severe fatique, syncope and epistaxis. On the physical examination, multiple telangiectases were found at the oral mucous membrane and tongue, cyanosis of the lips and fingers. The family history showed that his father have also had frequent episodes of epistaxis. Laboratory evaluation was notable for a haemoglobin value of 13 g/dL and hematocrit value 47%. Room air arterial blood gas values were pH 7.43, PO_2 52.2 mmHg, PCO_2 35.2 mmHg and oxygen saturation was 88%. Pulmonary function studies were normal. Chest radiograph showed a fairly well defined lesion in the left lung. On computed tomography (CT) scan and pulmonary angiography, multiple lesions were observed in the left lung (Figure 1, 2). Computed tomography of the brain was normal. We performed a left thoracotomy. Inferior pulmonary vein and pulmonary artery of the lower lobe were mobilised. Although the vessels of the lower lobe had been ligatured, hemorrhage and pulsation of the lesion continued. The bronchial artery was larger than normal size. Pulmonary arteriovenous aneurysm was supplied from both pulmonary and bronchial arteries. Because of the large fistula connection to the inferior pulmonary vein, local resection was not appropriate. Therefore lobectomy was performed. Wedge resection was performed for small lesion in the superior lingulary segment. Postoperative arterial blood gas values were pH 7.47, PO_2 92.2 mmHg, PCO_2 39.1 mmHg and oxygen saturation was 96%. The postoperative course was uneventful and he was discharged on the 10th postoperative day without any complication. He was asymptomatic and the other laboratory evaluation was normal after tree years.

Discussion

Pulmonary arteriovenous aneurysm may occur as an isolated

Adres: *Dr. Alpay Sarper, Akdeniz Üniversitesi Tıp Fakültesi, Göğüs Cerrahisi Ana Bilim Dalı, Antalya*
e-mail: *asarper@akdeniz.edu.tr*

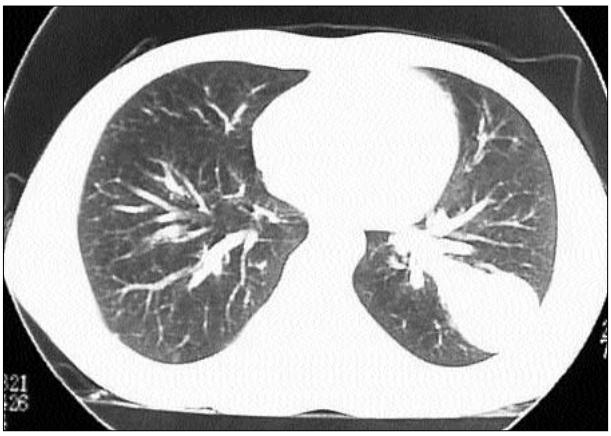

Figure 1. CT scan, showing the biggest lesion was in 4x5 cm, and located in the posteriobasal segment of the left lower lobe and connected by blood vessels to the hilus.

entity but is commonly associated with the syndrome of HHT, also known as ROWD. This disease is an autosomal dominant disorder characterised by mucocutaneous and visceral telangiectasis associated with recurrent episodes of epistaxis and gastrointestinal haemorrhage. The incidence of PAVA in HHT ranges from 7% to 15%, whereas HHT is an associated finding in approximately 35% of patients with PAVA [1,2]. Clinical presentation of PAVA ranges from an incidental finding on a chest roentgenogram in an asymptomatic patient (13%-56%) to polycythemia, cyanosis, congestive heart failure, and major neurologic deficits caused by paradoxical embolism. The most common complaints on presentation include dyspnea on exertion, palpitations, hemoptysis, or chest pain. Epistaxis, hematuria, or neurologic symptoms should alert the clinician to the possibility of coexisting ROWD. In large PAVA, the associated hypoxia and secondary polycythemia may give rise to neurologic symptoms and sings including headaches, confusion, dizziness, syncope, and cerebral vascular accidents, like our case.

Pulmonary arteriovenous aneurysm that has a systemic blood supply are rare, and it has the same hemodynamic consequences as arteriovenous fistulas of the general circulation have [1-3]. The bronchial arteries, internal mammary arteries, or aorta are the primary and immediate sources of the systemic arterial blood [2,5]. In our case, the fistula was supplied from pulmonary and bronchial arteries. We realized it during the operation, and this condition did not prevent to success of the operation. Chest radiographs are abnormal in approximately 98% of patients with PAVA. A single peripheral circumscribed noncalcified lesion connected by blood vessels to the hilus of the lung is the most common finding. Computed tomography scan usually demonstrates the lesion sufficiently well to be diagnostic. Angiography, however, is more reliable in the analysis of the angioarchitecture and is a necessary follow-up of the CT scan in those patients who are undergone interventional management of the fistulas [1,2,4].

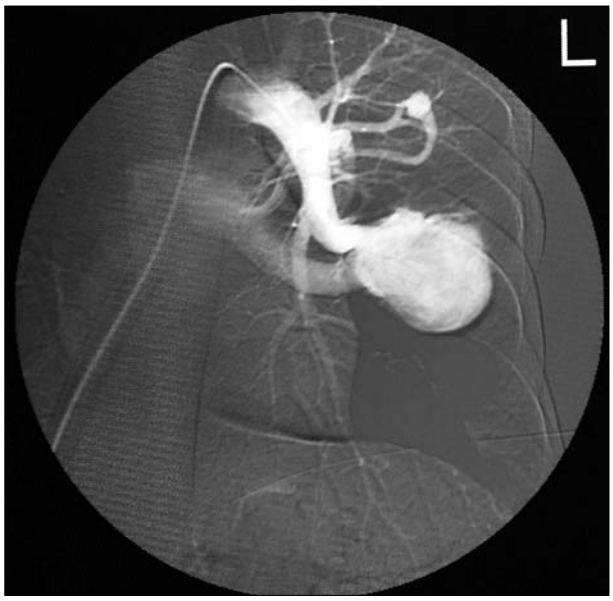

Figure 2. Pulmonary angiography showed that the lesion was pulmonary arteriovenous aneurysm and the large fistula connected directly to the inferior pulmonary vein.

Dines and co-workers [3] reported 11% mortality, and 26% morbidity rate in untreated patients followed for a mean of 6 years. Currently, most authors recommend that when lesions are symptomatic and enlarging or associated with complications, they should be treated [2]. Surgical excision of an isolated, single pulmonary arteriovenous fistula is successful, with minimal mortality and morbidity and little chance of recurrence of the lesion [2]. Because most fistulas are located subpleurally, they can be removed with conservative local resections. But like our case, same fistulas are located centrally, too large, and connected great vessels. Thus lobectomy may be required. However, patients unfit for surgery or those with multiple and bilateral lesions posed a difficult therapeutic problem. Selective radiographically guided embolization of multiple pulmonary arteriovenous fistulas that are unsuitable for surgical resection has proved to be a valuable therapeutic modality [1,2,6].

References

1. Burke CM, Safai C, Nelson DP, Raffin TA. Pulmonary arteriovenous malformations: A critical update. Am Rev Respir Dis 1986;134:334-9.
2. Shields TW. Congenital vascular lesions of the lungs. In: Shields TW, Cicero JL, Ponn RB, eds. General Thoracic Surgery. Philadelphia: Lippincott Williams and Wilkins, 2000:975-87.
3. Dines DE, Seward JB, Bernatz PE. Pulmonary arteriovenous fistula. Mayo Clin Proc 1983;58:176-81.
4. Hodgson CH, Burchell HB, Good CA, Claggett OT. Hereditary hemorrhagic telangiectasia and pulmonary

arteriovenous fistula: Study of a large family. N Engl J Med 1959;26:625-36.

5. Wispelaere JF, Trigaux JP, Weynants P, Delos M, Coene BD. Systemic supply to a pulmonary arteriovenous malformation: Potential explanation for recurrence. Cardiovasc Intervent Radiol 1996:19;285-7.

6. White RI, Lynch-Nyhan A, Terry P, et al. Pulmonary arteriovenous malformations: Techniques and long-term outcome of embolotherapy. Radiology 1988;169:663-9.