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

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

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Özet


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

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Introduction

Connections of the pulmonary artery to the pulmonary vein resulting in aneurysmatic dilatation are defined as pulmonary arteriovenous aneurysm (PVA) 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 PVA in 1897, and ROWD was set forth in 1901 by Osler [1]. We reported a rare case of ROWD associated with PVA, which received blood supply from the bronchial artery.

Case

A 20-year-old man was admitted to the hospital with hemoptysis, epistaxis and severe fatigue. 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 fistula


Discussion

Pulmonary arteriovenous aneurysm may occur as an isolated
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].

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

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