Giant Sporadic Pulmonary Arteriovenous Malformation

Mehmet Guli Cetincakmak1, Ozlem Abakay2, Abdurrahman Abakay2, Hadice Selimoglu Sen2, Abdullah Cetin Tanrikulu2

ABSTRACT

Pulmonary arteriovenous malformation is a rare clinical entity that can lead to emergency clinical situations such as bleeding, pneumothoraks and the long-term chronic hypoxic problems due to right-to-left shunt. Although pulmonary arteriovenous malformation is usually combined with hereditary hemorrhagic telangiectasia, it rarely seen as sporadic. Twenty-years-old female patient had consolidated areas in the right lung on chest X-ray. The multiple, inter-related vascular masses with early arterial intense contrast enhancement which the largest of them approximately 25x20 mm in size were observed in computed tomography. There were not a similar disease in family and epistaxis complaint, in patient’s medical history. A case of giant pulmonary arteriovenous malformation is presented with radiological findings and the literature data.

Key words: Arteriovenous malformation, lung, giant, sporadic

Introduction

Pulmonary arteriovenous malformation (PAVM) is a rare clinical entity that can lead to emergency clinical situations such as bleeding, pneumothoraks and the long-term chronic hypoxic problems due to right-to-left shunt (1,2). Although PAVM is usually combined with hereditary hemorrhagic telangiectasia, it rarely seen as sporadic (1). A case of PAVM is presented with radiological findings and the literature data.
Case

Twenty years-old female patient, was admitted with complaint of shortness of breath that ongoing about 3-4 years and increasing with effort. The patient’s dyspnea has increased in the last 1 month and consisted of at rest now. The patient have not cough and night sweats complaints. She had consolidated areas in the right lung on chest X-ray. The multiple, inter-related vascular masses with early arterial intense contrast enhancement which the largest of them approximately 25x20 mm in size were observed in lower lobe of right lung on thoracic computed tomography (CT) (Figure 1-4). The venous drainage of the masses was provided by pulmonary vein. There were not a similar disease in family and epistaxis complaint, in patient’s medical history. The dermatological examination was normal. Doppler echocardiographic examination was normal. The laboratory parameters were hematocrit 46.7 %, arterial blood mO2Hb: 92 %, mPCO2: 29.8 mm Hg, mPO2: 70.3 mmHg, respectively. The peripheral oxygen saturation was around 85% during rest. The patient was considered as a sporadic case.

Discussion

A pulmonary arteriovenous malformations (PAVM) is a rare vascular anomaly of the lung. Most cases tend to be simple AVMs (single feeding artery) although up to 20% of cases can have complex (two or more) feeding vessels (3). Symptoms of PAVM are usually based on the physiology of the right-to-left shunt. The blood passes from pulmonary artery to the pulmonary vein directly without capillary filtration and oxygenation. Cerebrovascular accident, transient ischemic attack, and brain abscesses may occur in this patients. (1,4). Convulsions, hemothorax, hemoptysis, and pneumotoraks may be seen in addition (2,5,6). More than 60 % of patients has been associated with an autosomal dominant hereditary hemorrhagic telangiectasia known as Osler-Weber-Rendu disease that characterized by epistaxis, mucocutaneous and visceral telangiectasia with a family history (1,7). Pulmonary arteriovenous malformations can be seen in the form of curvilinear lines around the mass with the drainage vein extending to the hilar region, on chest radiography. Contrast echocardiography, pulmonary perfusion scintigraphy, magnetic resonance imaging and pulmonary angiography can be used in diagnosis, although thoracic CT is the most useful method and gives more information about the anatomy of the mass. (4,8).

CT is often the diagnostic imaging modality of choice. The characteristic presentation of a PAVM on non-contrast CT is a homogeneous, well-circumscribed, non-calcified nodule up to several centimeters in diameter or the presence of a serpiginous mass connected with blood vessels (9). Occasionally associated phleboliths may be seen as calcifications. Contrast injection demonstrates enhancement of the feeding artery, the aneurysmal
part, and the draining vein on early-phase sequences. Sensitivity and specificity of PAVM detection in contrast-enhanced 16-detector CT has been reported at 83% and 78%, which compares favorably with digital subtraction pulmonary angiography of 70 and 100% (10).

Three-dimensional (3-D) helical CT is a technique in which CT data are collected continuously by a helical CT scanner; the CT window setting can be adjusted to preferentially visualize the vascular structures, which can be viewed from any angle as a 3-D shaded-surface display. In a study of 33 consecutive patients with 37 PAVM, noncontrasted 3-D helical CT scanning allowed full analysis of 76% of PAVM, compared with only 32% with unilateral pulmonary angiography (11). Unilateral angiography was often limited because of superimposed vessels. Concomitant analysis of 3-D helical images with cross-sectional images improved accuracy to 95%. Hyper-selective pulmonary angiography allowed analysis of 100% of PAVM but required additional contrast material and radiation (11).

Surgery was the only option in the treatment of pulmonary arteriovenous malformations in the past. However, transarterial embolization is usable as an effective method for PAVM treatment today (1,4,6,12). The studies are reporting recanalization and the need for re-treatment in 5-17% ratio after transarterial embolization, in the literature. The surgical treatment was recommended due to the multiple and inter-related lesions, in our patient. The patient was admitted to the cardiology department due to the development of preoperative cardiac arrhythmias. The patient was not followed by us later on.

Hereditary hemorrhagic telangiectasia should be investigated in patients with PAVM and transarterial embolization should be considered as first-line treatment. The surgery is another treatment option when embolization is not possible.

References