CASE REPORT

A pulmonary arteriovenous malformation treated with percutaneous intervention

Perkütan girişim ile tedavi edilen pulmoner arteriyovenöz malformasyon olgusu

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Summary– A pulmonary arteriovenous malformation (PAVM) is a rare anomaly that may have significant clinical complications. PAVMs are commonly seen in patients with hereditary hemorrhagic telangiectasia, while some 10% of PAVMs may be idiopathic. PAVMs can cause cyanosis, fatigue, polycythemia, and paradoxical thromboembolic complications. The diagnosis and treatment of a PAVM should be performed with great care, as the disorder may be fatal if not properly treated. Percutaneous closure (such as embolization) can be very beneficial. Presently described is the case of a 23-year-old man with an idiopathic PAVM who was treated percutaneously with 3 vascular plugs.

Özet– Pulmoner arteriyovenöz malformasyonlar (PAVM), klinik komplikasyonlara neden olan ve nadir görülen anomalilerdir. PAVM'ler, kalıtsal hemorajik telenjektazisi olan hastalarda sık görülür. PAVM'lerin yüzde onu idiyopatik olabilir. PAVM'ler siyanoz, yorgunluk, polisitemi ve paradoksik tromboembolik komplikasyonlara neden olabilir. PAVM'nin tanısı ve tedavisi çok dikkatli yapılmalıdır. Bu hastalık uygun şekilde tedavi edilmezse ölümcül olabilir. Günümüzde perkütan kapama (embolizasyon ile) tedavide büyük önem taşımaktadır. Bu yazıda, idiyopatik PAVM'si olan ve üç vasküler tıkaç ile perkütan tedavi ettiğimiz 23 yaşındaki erkek hasta sunuldu.

A pulmonary arteriovenous malformation (PAVM) is a rare anomaly with common presentation complaints, such as dyspnea on exertion, nasal bleeding, fatigue, bluish discoloration (lips and nail beds), and hemoptysis. These arteriovenous malformations are communication between the pulmonary artery and vein.[1] PAVMs are associated with hereditary hemorrhagic telangiectasia (HHT) in up to 85% of cases, whereas the remaining 10% to 15% are sporadic fistulas. Some 15% to 35% of HHT patients present with a PAVM.[2] It can cause severe morbidity and mortality as a result of complications like a cerebral vascular accident, brain abscess, hemothorax, or life-threatening hemoptysis if not treated early and adequately.[3]

CASE REPORT

A 23-year-old male patient presented at the clinic with complaints of fatigue and shortness of breath. A physical examination revealed clubbing (Fig. 1a). A posterior-anterior lung X-ray illustrated opacity in the right paracardiac area (Fig. 1b). The level of peripheral arterial oxygen saturation recorded was 90%, the hemoglobin value was 18.9 g/dL and the hematocrit result was 56.8%. A transthoracic echocardiography examination indicated that the left ventricle systolic and end-diastolic dimensions were 33 to 53 mm, the right atrium was 30 mm, the right ventricle 32 mm, and the systolic pulmonary artery pressure 30 mm Hg. According to cardiac catheterization, the oxygen saturation was 69% in the vena cava superior, 80% in the vena cava inferior, 72% in the right atrium and right ventricle, 75% in the pulmonary artery, 90% in the right pulmonary vein, 98% in the left pulmonary vein, 92% in the left ventricle, and 92% in the ascending aorta. Pul-

Abbreviations:

HHT	*Hereditary hemorrhagic telangiectasia*
PAVM	*Pulmonary arteriovenous malformation*

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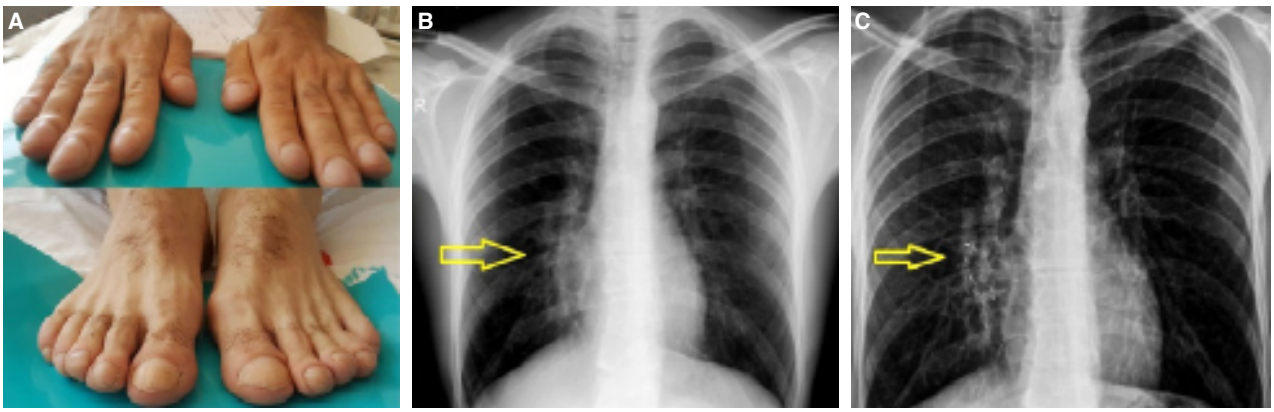

Figure 1. **(A)** Clubbing in the hands and feet; **(B)** Right paracardiac opacity in a posterior-anterior lung X-ray; **(C)** Right paracardiac vascular plugs and decreased opacity in a posterior-anterior lung X-ray.

monary multislice computerized tomography showed multiple PAVMs at the inferior lobe of right lung (Fig. 2a, b). Pulmonary angiography illustrated complex PAVMs (Fig. 3a, Video 1*). The PAVMs of the inferior lobe of the right lung were occluded with 3 plugs (Amplatzer Vascular Plug II, 18 mm, 10 mm, 8 mm; St Jude Medical, Inc., St. Paul, MN, USA) (Fig. 3b). Unfractionated heparin treatment and endocarditis prophylaxis were administered prior to the procedure. Subsequently, the arteriovenous flow decreased (Fig. 3c, 1c, Video 2*).

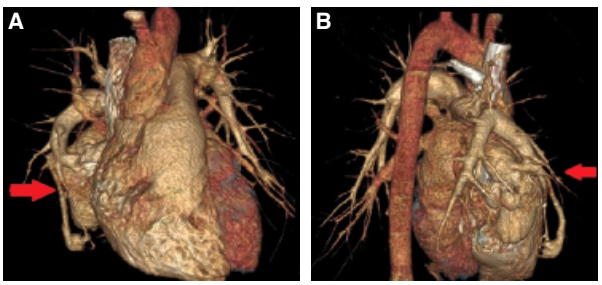

Figure 2. **(A)** Anterior view of pulmonary arteriovenous malformations (PAVMs) at the right lung seen on a pulmonary multislice computerized tomography image; **(B)** Posterior view of PAVMs at the right lung observed with pulmonary multislice computerized tomography.

The patient was discharged in good clinical condition with a prescription for clopidogrel 75 mg/day and acetylsalicylic acid 100 mg/day for 6 months. If a later interventional or surgical procedure was performed, the patient was advised that endocarditis prophylaxis be administered.

DISCUSSION

PAVM is a rare anomaly. According to a study conducted by Sloan and Cooley in 1953, only 3 cases

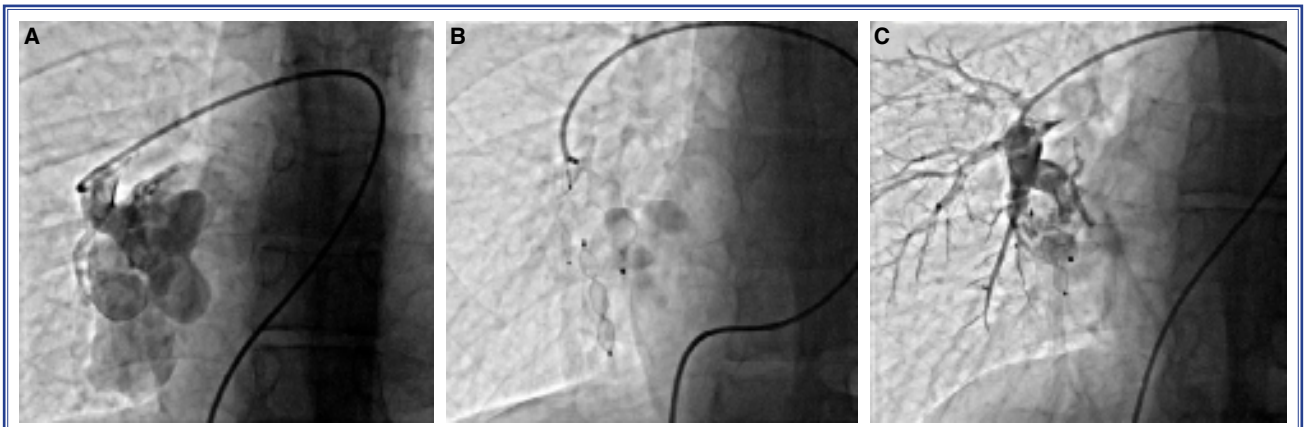

Figure 3. **(A)** Complex pulmonary arteriovenous malformations (PAVMs) as seen with pulmonary angiography; **(B)** Fluoroscopic view of 3 vascular plugs in the inferior lobe of the right lung; **(C)** Fluoroscopic view of the decreased arteriovenous flow in the PAVMs after the plug insertion.

of PAVM were detected in 15,000 consecutive autopsies.[4] PAVMs are anatomically classified as focal, complex, or diffuse. The focal type is the most common. It has a single segmental feeding artery, though it may have multiple subsegmental branches, whereas the complex type has multiple segmental feeding arteries (~20%). Examples of the diffuse variation are rare (~5%). This type has a combination of simple and complex malformations within a diffuse lesion.[5]

PAVMs are etiologically divided into 4 types. The most common is related to HHT (~85%), an autosomal dominant disorder in which vascular malformations are frequently seen in the skin, nasopharynx, gastrointestinal tract, lungs, and brain. HHT typically consists of a triad of cutaneous telengiectasia, recurrent epistaxis, and a family history of this disorder.[6] PAVMs occur twice as often in female patients.[6] Most often, idiopathic congenital PAVMs (~15%) are single lesions. They are less likely to become enlarged, and can be successfully treated with embolotherapy. PAVMs may also be acquired secondary to a chronic infection (~<1%), such as schistosomiasis, actinomycosis, tuberculosis, liver cirrhosis, or metastatic thyroid cancer. PAVMs also may be iatrogenic (~<1%) due to cardiovascular intervention or congenital cyanotic heart disease.[7]

In a PAVM, blood bypasses the pulmonary capillary bed where normal oxygen exchange takes place, leading to desaturated blood returning to the pulmonary veins. When the shunting becomes significant, cyanosis may occur, and consequently, the concentration in hemoglobin and the hematocrit increases. PAVMs are more common in the lower lobes, which may lead to orthodeoxia and platypnea in advanced cases. Bleeding from the nose and gastrointestinal telangiectasias may reduce hemoglobin in HHT, potentially causing anemia and stroke due to paradoxical embolization. Mortality is higher in cases of bilateral, diffuse PAVMs.[8]

There are several methods available to measure the right-to-left shunt; however, the level of sensitivity and specificity are not high. If the pulmonary artery oxygen saturation level is less than 85% or the arterial oxygen saturation level is less than 96%, it suggests that the potential shunt fraction may be greater than 5%. In the present case, the pulmonary artery oxygen saturation value was 75% and the peripheral arterial oxygen saturation was 90%, an indication that our patient's shunt fraction was greater than 5%.

Though the presence of dyspnea, cyanosis, and clubbing is described in textbooks as the triad for PAVM, the combination was unequivocally present in only 10% of patients with PAVM in 1 study.[9,10] Approximately 72% of patients with PAVM have symptoms referable to PAVM or underlying HHT. Symptoms related to PAVM often develop between the fourth and sixth decades of life, when the shunted blood exceeds 25% of total blood volume.[4,11] Dyspnea on exertion is the most common complaint in patients with PAVM, and it is seen in about half of patients (10). Our patient complained of dyspnea and fatigue. Other symptoms attributable to PAVM include hemoptysis (10%), chest pain (6%), finger clubbing (20%), cyanosis (18%), and thoracic murmurs (3%).[10]

The natural course of PAVM is not benign. These lesions can be associated with a variety of life-threatening complications, such as stroke, brain abscess, hemothorax, and hemoptysis, especially in women.[3] Rupture of a PAVM can occur at any age, independent of lesion size.[12] Without appropriate treatment, mortality exceeds 11%.[13]

All symptomatic PAVMs, as well as asymptomatic PAVMs that are larger than 2 cm in size or have feeding arteries larger than 2 mm, should be treated with surgery because of the risk of paradoxical embolism.[14] The treatment of choice in patients with multiple or bilateral PAVMs is transcatheter embolotherapy with balloons or stainless steel coils, or vascular plugs. Embolotherapy can be performed with only 1 coil in a small PAVM. In large and high flow PAVMs, a coil and balloon combination or a vascular plug treatment can be applied. In a large and high-flow PAVM, the coil could be embolized to a venous site or systemic circulation. The technique of coil embolotherapy requires selective catheterization of all feeding arteries of the PAVM, and then the catheter tip is positioned as close to the neck of the PAVM as possible. The coil is advanced through the catheter and released at the proper position, angiography is repeated, and additional coils are positioned if needed until blood flow to the PAVM has stopped. In a combination approach, a balloon and a delivery catheter are advanced through the feeding artery of the PAVM. The flow is halted with the balloon, the coil is advanced through

the delivery catheter and released at the proper position using flow control. Large PAVMs are usually treated with vascular plugs. The delivery catheter tip is advanced to the proper position, the vascular plug is advanced through the catheter and released at the proper location. To prevent paradoxical embolization, the device should have a diameter 2 mm greater than the diameter of the feeding vessel of the PAVM when using coils, or a diameter 20% to 30% larger than the feeding vessel of the PAVM when using plugs. The procedures are performed with heparinization and endocarditis prophylaxis. Embolization therapy eliminates the need for surgical intervention and has lower rates of morbidity and mortality.[8] Follow-up with multiple detector computed tomography at 6 to 12 months must be performed to determine reperfusion of the PAVM and opening of an occult PAVM.[15]

Conclusion

A PAVM is a rare disorder that may be present in patients with very common presentations. A detailed history and a high degree of suspicion are of immense importance for a final diagnosis. Percutaneous embolization has become the treatment of choice of PAVM due to its safety and efficacy in experienced hands.

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**Supplementary video file associated with this article can be found in the online version of the journal.*

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Anahtar sözcükler: Perkütan tedavi; pulmoner arteriyovenöz malformasyon; vasküler tıkaç.