

Thrombosed giant proximal pulmonary artery aneurysm

Trombozlu dev proksimal pulmoner arter anevrizması

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Bu yazıda, 22 yıl önce tanı konan Eisenmenger sendromu zemininde dev proksimal pulmoner arter anevrizması gelişen ve durumun trombüs oluşumu ile daha ciddileştiği 36 yaşında bir erkek hasta sunuldu. Hastada son bir aydır paroksizmal atriyal fibrilasyon atakları gelişmişti. Hastanın fonksiyonel kapasitesi New York Kalp Birliği sınıf III idi. Akciğer grafisinde sol pulmoner arterde anevrizmatik genişleme görüldü. Transtorasik ekokardiyografi ve çokkesitli bilgisayarlı tomografi ile değerlendirilen hastada kalın, kalsifik yapıdaki pulmoner kapakta hafif darlık (tepe sistolik gradiyent 35 mmHg) ve orta derecede yetersizlik saptandı. Ortalama pulmoner arter basıncı 50 mmHg olarak hesaplandı. Ana pulmoner arter, sol pulmoner arter ve sağ pulmoner arter çapları sırasıyla 6.5 cm, 10 cm ve 3.7 cm ölçüldü. Anevrizmatik olan sol pulmoner arterin lümeni yoğun trombüs ile doluydu. Hasta, pulmoner arter rekonstrüksiyonu ve kalp-akciğer nakli için kardiyovasküler cerrahi kliniğine yönlendirildi. Ayrıca, trombüs ve paroksizmal atriyal fibrilasyona yönelik warfarin, atriyal fibrilasyona yönelik metoprolol ve pulmoner hipertansiyona yönelik bosentan tedavisine bağlandı. Tedavinin ilk ayından sonra hastanın fonksiyonel kapasitesinde düzelmeye başladı. Bu tedavinin sürdürüldüğü bir yıl içinde komplikasyon görülmedi.

Anahtar sözcükler: Anevrizma/komplikasyon; Eisenmenger kompleksi/komplikasyon; hipertansiyon, pulmoner; pulmoner embolizm/etioloji; bilgisayarlı tomografi.

Pulmonary artery aneurysm (PAA) is defined as pulmonary artery dilatation of more than 4 cm. Aneurysms in the main pulmonary artery or both branches are classified as proximal PAA.^[1] Pulmonary artery aneurysms are very rare. Deterling and Clagett^[2] found eight cases with PAA (0.0073%) in 109,571 autopsies. Pulmonary hypertension is the major etiological factor in the development of pulmonary artery aneurysm.^[3] In this

We present a 36-year-old male patient with a previous diagnosis (22 years) of Eisenmenger's syndrome, who had a giant proximal pulmonary artery aneurysm complicated by massive thrombus formation. The patient had been experiencing paroxysmal atrial fibrillation attacks for the past month. His functional capacity was of New York Heart Association class III. Chest radiography showed aneurysmal dilatation in the left pulmonary artery. The patient was assessed by transthoracic echocardiography and multislice computed tomography. There was mild narrowing in the thick and calcified pulmonary valve (peak systolic gradient 35 mmHg) and moderate regurgitation. The mean pulmonary artery pressure was estimated at 50 mmHg. The diameters of the main, left, and right pulmonary arteries were 6.5 cm, 10 cm, and 3.7 cm, respectively. There was a massive thrombus in the aneurysmal left pulmonary artery. The patient was referred to the cardiovascular surgery department for pulmonary artery reconstruction and cardiopulmonary transplantation. In addition, medical treatment was instituted with warfarin for thrombus and paroxysmal atrial fibrillation, metoprolol for atrial fibrillation, and bosentan for pulmonary hypertension. The patient's functional capacity showed improvement after the first month of medical treatment and no complications were seen within a year follow-up.

Key words: Aneurysm/complications; Eisenmenger complex/complications; hypertension, pulmonary; pulmonary embolism/etiology; tomography, X-ray computed.

study, we presented a patient with a previous diagnosis of Eisenmenger's syndrome, who had a giant proximal pulmonary artery aneurysm complicated by massive thrombus formation.

CASE REPORT

A 36-year-old male patient who was diagnosed with Eisenmenger's syndrome 22 years ago was referred to our

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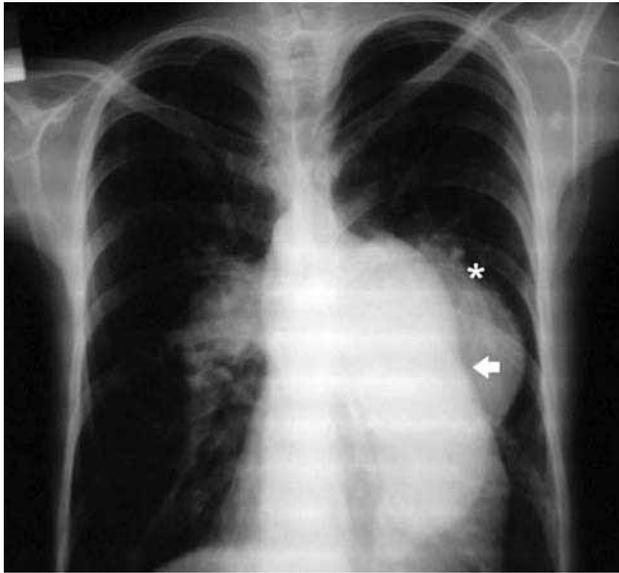


Figure 1. Chest X-ray image of an enlarged pulmonary conus.

laboratory clinic for echocardiography. The patient had been experiencing paroxysmal atrial fibrillation attacks for the past month. His functional capacity was of New York Heart Association class III. The patient had central cyanosis and clubbing of the fingers. His blood pressure was 105/60 mmHg, and pulse rate was 80/min. Cardiac examination revealed an apex beat on the left side, rhythmic heart sounds, and a strong S2 sound. A 2/6 early diastolic murmur at the upper sternal border and a 2/6 pansystolic murmur at the lower sternal border were heard. Electrocardiography showed a sinus rhythm, loss of R-wave progression in leads V1-V3 and strain pattern and negative T-wave in leads V5-V6 were found. Chest radiography demonstrated a gastric air chamber on the right and heart apex on the left. In addition, increased

cardiothoracic ratio, right hilar fullness, enlarged pulmonary conus, and aneurysmal dilatation in the left pulmonary artery were found on the chest x-ray (Figure 1). His complete blood count showed a hemoglobin level of 15.4 g/dL, leukocyte count of 7900/mm³, and platelet count of 145,000/mm³. His blood biochemistry test results and thyroid function tests were normal. Arterial blood gas analysis showed a pH value of 7.4, pO₂ of 46.4 mmHg, pCO₂ of 32.4 mmHg, HCO₃ of 21.4 mmol/L, and O₂ saturation of 83.6%.

The patient with isolated levocardia was assessed by transthoracic echocardiography followed by multislice computed tomography (CT) (Toshiba Aquilion 16, Japan) to perform detailed visualization of cardiac anatomy. Images showed right atrium positioned on the right and the left atrium on the left. Large, sinus venosus-type atrial septal defect in interatrial septum was observed. The right atrium opened into the morphological left ventricle via the thick dysplastic mitral valve, while the left atrium opened into the morphological right ventricle which was more trabecular with a moderator band, via tricuspid valve (atrioventricular inconsistency) (Figure 2a). There was also moderate regurgitation of the mitral and tricuspid valves. Pulmonary artery arose from morphological left ventricle, while aorta arose from morphological right ventricle (ventriculoarterial inconsistency).

Atrioventricular and ventriculoarterial connections were consistent with corrected transpositions of the great arteries. The systolic functions of the morphological right ventricle, functioning as a systemic ventricle was within the normal range. In the basal region of intraventricular septum, a 2 cm perimembranous ventricular septal defect was detected. Aortic valve was on the right next to pulmonary valve (Figure 2b). A mild narrowing (peak

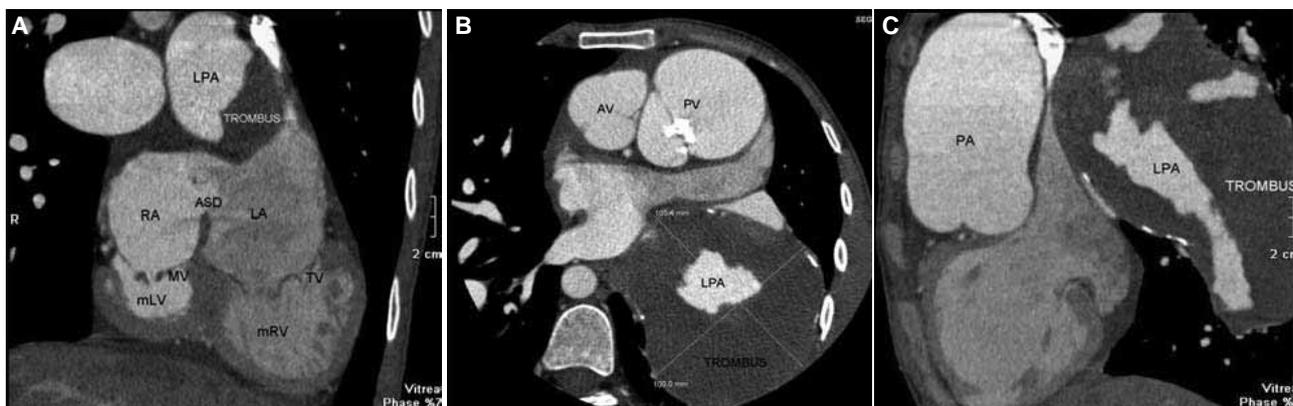


Figure 2. Multislice computed tomography images: (A) Large sinus venosus-type atrial septal defect and thrombus in the lumen of the left pulmonary artery; (B) calcified pulmonary valve and massive thrombus in the lumen of the aneurysmatic left pulmonary artery; (C) aneurysmatic main pulmonary artery and aneurysmatic left pulmonary artery with massive thrombus in the lumen. ASD: Atrial septal defect; LPA: Left pulmonary artery; PA: Pulmonary artery; MV: Mitral valve; mLV: Morphologic left ventricle; TV: Tricuspid valve; mRV: Morphologic right ventricle; AV: Aortic valve; PV: Pulmonary valve.

systolic gradient of 35 mmHg) and moderate regurgitation were found in the thick and calcified pulmonary valve. Mean pulmonary artery pressure was 50 mmHg as assessed by continuous flow Doppler through the pulmonary regurgitation jet. The main pulmonary artery and branches (particular left pulmonary artery) were markedly dilated. Multislice CT demonstrated main pulmonary artery of 6.5 cm, left pulmonary artery of 10 cm, and right pulmonary artery of 3.7 cm. There was a massive in situ thrombus in the lumen of the aneurismatic left pulmonary artery (Figure 2c).

The patient was referred to the cardiovascular surgery department for pulmonary artery reconstruction and cardiopulmonary transplantation. In addition, medical treatment was instituted with warfarin for the pulmonary artery thrombus and paroxysmal atrial fibrillation, metoprolol 50 mg/day for rate control in case of any recurrent atrial fibrillation, and bosentan 62.5 mg bid for pulmonary hypertension. The patient's functional capacity showed improvement after the first month of medical treatment with bosentan and no complication has been seen within the past one year of follow-up.

DISCUSSION

In this paper, a patient with a previous diagnosis of Eisenmenger's syndrome, who had a giant proximal pulmonary artery aneurysm complicated by massive in situ thrombus formation, has been presented. Although an aneurysm of 15 cm in the main pulmonary artery has been reported in literature,^[4] our case was the first to report aneurysm involving any of the branches of the pulmonary artery and approaching 10 cm in diameter. As mentioned earlier, pulmonary hypertension is significant in the development of PAA. Other etiologic factors include congenital cardiac anomalies with left-to-right shunt, congenital absence of the pulmonary valve, connective tissue diseases (Marfan syndrome, Behcet's disease), idiopathic causes (isolated Hughes-Stovin syndrome), infections, trauma, and neoplasia.^[5,6] In our case, ventricular and atrial septal defects with left-to-right shunt increased the volume and pressure load on the pulmonary artery circulation, and pulmonary hypertension which developed later increased hemodynamic tension, resulting in PAA. On the other hand, presence of a giant aneurysm of 10 cm in our patient may suggest an existing internal disorder on the wall of the vessel.^[7] In addition, the changed flow dynamics due to calcified and narrow pulmonary valve and of the additional volume load due to pulmonary insufficiency might also have contributed to the development of the giant aneurysm. Elastic fiber breakdown in the media of the pulmonary artery with

increased ground substance was reported in patients with congenital pulmonary valve disorder.^[8] Dilatation of the pulmonary artery is not always associated with the extent of the stenosis in the pulmonary valve.^[9] This mechanism is similar to dilatation of the aortic root and ascending aorta presenting with no significant stenosis or regurgitation of the valve in patients with bicuspid aortic valve.^[10,11]

Complications associated with pulmonary artery aneurysm include bronchial pressure, pulmonary artery dissection, pulmonary artery laceration, and thrombus formation as seen in our case.^[1] About 20-30% of the patients with Eisenmenger's syndrome present with pulmonary artery thrombus.^[12,13] Flow decrease in the aneurysm and endothelial dysfunction are predisposing factors for thrombus formation. There are a limited number of studies evaluating the therapeutic and preventive effects of anticoagulation on thrombus formation. In these patients some authors recommend using warfarin treatment under closed observation for patients without hemoptysis or just mild hemoptysis.^[14,15] As our patient did not experience hemoptysis, we initiated warfarin treatment to melt pulmonary artery thrombus and prevent systemic embolization in paroxysmal atrial fibrillation.

No consensus has been reached on the treatment of pulmonary artery aneurysm. However, low-pressure aneurysms without pulmonary hypertension are considered as benign. Veldtman et al.[8] recommended scheduling surgical intervention based on changes in the size and function of the right ventricle due to pulmonary stenosis or regurgitation, and not the extent of the aneurysm in patients with low-pressure aneurysms. Surgical intervention is unquestionable under conditions of severe life threatening hemoptysis or pressure. As the risk of laceration and dissection is higher in patients with high-pressure pulmonary artery aneurysm with underlying pulmonary hypertension as seen in our case, aggressive surgical management is recommended.^[16] Accordingly, our patient was referred for pulmonary artery reconstruction and cardiopulmonary transplantation. It is noteworthy that this high-risk surgical intervention is performed only at specific centers. The patient with a NYHA class III functional capacity was instituted with bosentan, an endothelin receptor antagonist until surgery. The ability of bosentan in improving hemodynamics and exercise capacity in patients with Eisenmenger's syndrome has been demonstrated in clinical studies.^[17,18] We also observed improvement in the functional capacity of our patient following the first month of medical treatment with bosentan.

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