Pulmonary Arteriovenous Fistulas Without Hereditary Hemorrhagic Telangiectasia: Case Report

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HEREDİTER HEMORAJİK TELENJİEKTAZİNİN EŞLİK ETMEDİĞİ BİR ARTERİOVENÖZ FİSTÜL OLGUSU

ÖZET

Pulmoner arteriyovenöz fistüller nadir görülen bir anomalidir ve çoğunlukla herediter hemorajik telenjektazili hastalarda bulunur. Tanı konulmaması veya tedavi edilmemesi durumunda yaşamı tehdit edecek komplikasyonlara neden olabilir. Çoğunlukla herediter hemorajik telenjektazinin klinik bulguları göze çarpmaktadır. Bu yazıda bilateral pulmoner arteriyovenöz fistül saptanan ve herediter hemorajik telenjektazi bulunmayan bir olgu sunulmaktadır. ***Türk Kardiyol Dern Arş 2002; 30:***

Anahtar kelimeler: *Pulmoner arteriyovenöz fistül, sistolo-diyastolik üfürüm*

Pulmonary arteriovenous fistula occurs as a result of defective embriologic development of pulmonary capillary system [1]. As a result of right-to-left shunt, it causes hypoxemia and polycytemia. Although some patients are asymptomatic, most of the cases might develop dyspnea, cygnosis, clubbing and hemoptysis while some has neurologic symptoms due to extrapulmonary complications [2-5]. Epistaxis due to hemorrhage of telangiectasias in upper respiratory tract is the most common (79%) presenting symptom which is related to hereditary hemorrhagic telangiectasia accompanying pulmonary arteriovenous fistulas in 88% cases [2].

During the course of pregnancy, pulmonary arteriovenous fistulas can grow and lead to symptoms such as dyspnea and hypoxia. Some of the asymptomatic patients are first diagnosed during pregnancy presenting with life-threatening complications [3,6,7].

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Whatever the presenting symptoms are, 88% of the cases of pulmonary arteriovenous fistulas are associated with hereditary hemorrhagic telangiectasia. Herein, we report bilateral pulmonary arteriovenous fistulas in a case without hereditary hemorrhagic telangiectasia which is a rare presentation.

CASE REPORT

A 21 year old man admitted to hospital with dyspnea and chest pain.Physical examination showed; blood pressure of 110/70 mmHg, pulse of 70/min, clubbing at his fingers, systolo-diastolic murmur on left 6th intercostal space between anterior and mid axillary lines. Electrocardiogram was normal. Telecardiogram revealed ovoid opacities next to left paracardiac region (4.0x2.5 cm) and in apical segment of right lower lobe (5.0x3.2 cm) (Fig 1).

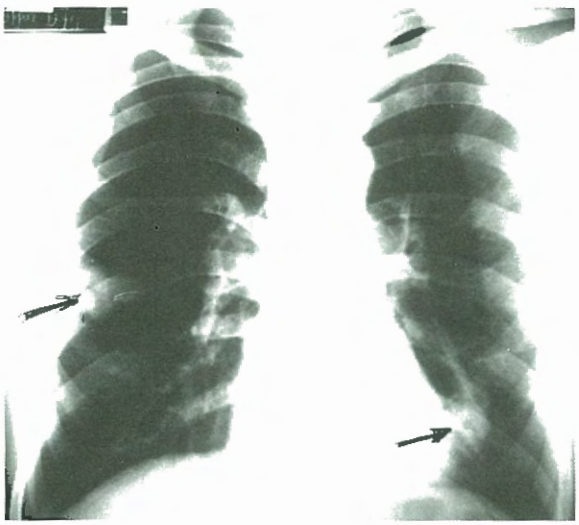
Arterial blood gas examination revealed hypoxemia and hypercapnia, PaO_2 and $PaCO_2$ were 86 and 57 mmHg, respectively while the patient was breathing air room and supine position. Other biochemical and hematologic parameters were within normal ranges. Computed tomographic examination of thorax showed bilateral lobulated soft-tissue densities compatible with arteriovenous malformations in right lower lobe and left lingular segment.

After the right and left catheterization, coronary arteriography, aortography (including carotid, mesenteric, renal and iliac angiograms) and left ventriculography were done and all were normal. Left pulmonary arteriogram showed an artery reaching to the lesion, filling-in an aneurysmal sac and a large draining vein at apical segment of the left lower lobe (Fig 2 A-B). Right pulmonary angiogram demonstrated peripherally located aneurysmal sac together with the feeding artery and the draining vein (Fig 3). Diagnosis of bilateral pulmonary arteriovenous fistula was obtained. Embolization recommended but the patient rejected.

DISCUSSION

Age, sex, clinical and laboratory presentation of our case is similar to previously reported cases except the finding of systolo-diastolic murmur heard on left 6th intercostal space in our case. Although a few ca-

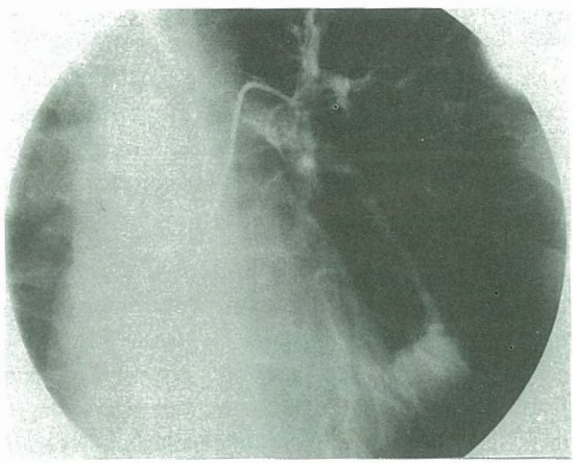
Figure 1. Antero-posterior telecardiogram shows bilateral pulmonary opacities (arrows).



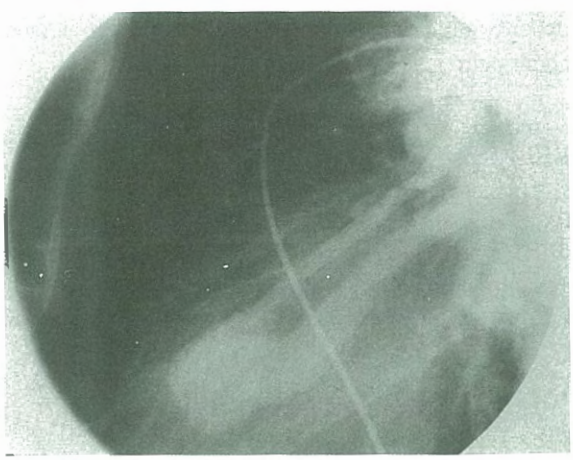
ses reported murmur or bruit as a presenting finding, the largest study reported by White et al [2] unfortunately did not expressed clinical findings in detail [6,7]. Though it was reported rarely, we do not believe that a bruit or murmur is a specific or rare finding for pulmonary arteriovenous fistulas. Location and multiplicity of the lesions in our case were both relevant to the past literature [2]. Although neurologic symptoms such as migraine (%43), transient ischemic attacks (%37), stroke (18%) and brain abscess (9%) reported previously, our patient had none of these symptoms and findings. Furthermore if the patient is left untreated, some of them will have maintain significant morbidity and mortality because of stroke, brain abscess, hemoptysis and hemothorax [8]. Epistaxis, which is the most common presenting symptom of HHT [2], was not present in our patient which made diagnosis uneasy.

Hereditary hemorragic telangiectasia (HHT) is present in 88% of the reported cases of pulmonary arteriovenous fistulas and pulmonary arteriovenous fistulas have been found in approximately 15% of patients with HHT [2-4]. White et al [2] reported 276 pulmonary arteriovenous fistulas in 76 patients which is the largest population of patients in the literature. Although the concurrence of HHT with pulmonary arteriovenous fistulas were reported as 30-50% in previous reports, they mentioned that with careful evaluation, HHT was found in 88% of their patients. HHT was not present in our patient which is a rarity of our case.

Figure 2. Left paracardiac arteriovenous fistula on left pulmonary angiogram
A. Antero-posterior position B. Left lateral position

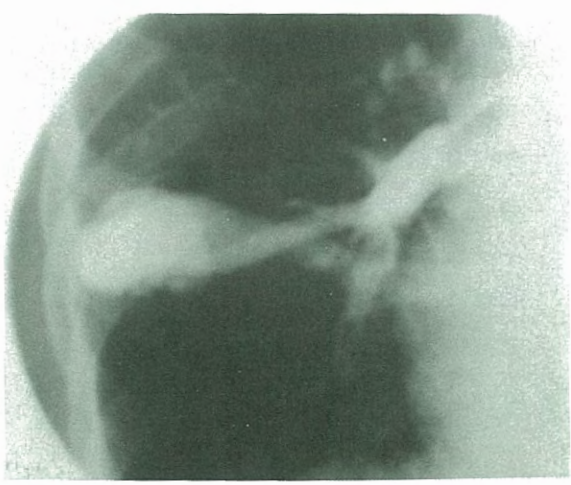


A



B

Figure 3. Arteriovenous fistula at right pulmonary angiography (antero-posterior position).



Until 1980's, the treatment was limited by surgical resection or ligation of the PAF [9]. In 1977, first Porstmann [10] and then Taylor [11] reported that coil embolization was effective treatment for PAF. Afterwards White et al [2], started to use balloon embolization therapy. Coil or balloon embolizations are still preferred as a first choice in the treatment of PAF. Embolization is proposed but rejected by our patient.

Early diagnosis of pulmonary arteriovenous fistulas is essential to prevent further life-threatening complications. Although it is a rare clinical presentation, we must keep in mind that pulmonary arteriovenous fistulas may present without hereditary hemorrhagic telangiectasia, as in our case. In conclusion, in cardiac examination the patients with systolo-diastolic murmur, clubbed fingers and abnormal pulmonary opacity in telecardiogram should alert the physicians about pulmonary arteriovenous fistula.

REFERENCES

1. Szilagyi DE, Elliott P, De Russo FJ, Smith FJ: Peripheral congenital arteriovenous fistulas. Surgery 1965: 57: 61-80

2. White RI, Lynch-Nyhan A, Terry P, Buescher PC, Farmlett EJ, Charnas L, et al: Pulmonary arteriovenous malformation: Techniques and long-term outcome of embolotherapy. Radiology 1988:169: 663-9

3. Gammon RB, Miksa AK, Keller FS: Osler-Weber-Rendau disease and pulmonary arteriovenous fistulas.: Chest 1990;90: 1522-24

4. Dines DE, Arms RA, Bernatz PE, Gomes MR: Pulmonary arteriovenous fistulas. Mayo Clin Proc 1974: 49: 460-5

5. Dines DE, Sevard JB, Bernatz PE: Pulmonary arteriovenous fistula. Mayo Clin Proct 1983: 58: 176-181

6. Swinburne AJ, Fedullo AJ, Gangemi R, Mijangos JA: Hereditary telengiectasia and multiple pulmonary arteriovenous fistulas. Chest 1986;89: 459-60

7. Swinburne AJ: Pregnancy and pulmonary arteriovenous fistula: NY State J Med 1992;92: 515-16

8. Burke CM, Safai C, Nelson DP, RaffiN TA: Pulmonary arteriovenous malformations: A critical update. Am Rev Respir Dis 1986:134: 334-39

9. Browns S, Wright PW, Renner JW, Riker JB: Staged bilateral thoracotomies for multiple pulmonary arteriovenous malformations complicating hereditary hemorrhagic telangiectasia. J Thorac Cardiovasc Surg 1982: 83: 285-89

10. Porstman W: Therapeutic embolization of arteriovenous fistula by catheter technique. In: Kelop O, ed. Current Concepts in Pediatric Radiology. Berlin Springers 1977: 23-31

11. Taylor BG, Cockerill EM, Manfredi F, Klatte EC: Therapeutic embolization of pulmonary artery in pulmonary arteriovenous fistula. Am J Med 1978: 54: 360-63