CASE REPORT

Çocukluk çağında siyanozun nadir bir sebebi: Pulmoner arteriyovenöz malformasyon

A rare cause of cyanosis in childhood: Pulmonary arteriovenous malformation

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Özet- Pulmoner arter ve pulmoner ven arasında anormal bağlantı olması olarak tanımlanan pulmoner arteriyovenöz malformasyon nadir görülen bir durumdur. Genellikle doğumsal olarak ortaya çıkmasına rağmen herediter he- morajik telenjiektazi ile birlikte de olabilir. Klinik bulgular, fistülün sayısı ve boyutu ile orantılı olarak şant miktarıyla değişmekte olup hastalarda siyanoz ve solunum sıkıntısı görülebilir. Hastalık tedavi edilmediği takdirde kalp yetersizliğine ve enfektif endokardite yol açabilir; anevrizmal fistül yırtılabilir. Anormal vasküler bağlantının transkate- ter embolizasyonu bu hastalığın yönetimindeki güncel tedavi yöntemidir. Bu makalede 8.5 yaşında bir çocuk hasta sunuldu. Hastada çabuk yorulma belirtisi kendini gösteriyordu. Transkutanöz oksijen satürasyonu %75 idi, incelemelerinde pulmoner arteriyovenöz malformasyon tespit edildi. Başarılı transkateter fistül embolizasyonu uygulandı.

Summary- Pulmonary arteriovenous malformation, which is defined as the presence of an abnormal connection between the pulmonary artery and pulmonary vein, is rarely seen. Although it generally presents as a congenital condition, it may be accompanied by hereditary hemorrhagic telangiectasia. Clinical signs vary according to the amount of shunt in proportion to the number and size of the fistulae. Patients may present with cyanosis and respiratory trouble. If the disease remains untreated, it may result in cardiac failure and infective endocarditis, thereby leading to the rupture of the aneurysmal fistula. Transcatheter embolization of abnormal vascular connection is the current treatment method in this disease. This article describes the case of an 8-year-old child. He was presented with the symptom of getting tired quickly. Transcutaneous oxygen saturation of 75%, and pulmonary arteriovenous malformation were detected in his examination. Successful transcatheter embolization of the fistula was performed.

Pulmonary arteriovenous malformation (pAVM) is the presence of direct abnormal, communication between pulmonary arteries and pulmonary veins without interconnecting capillaries. It is a rare pathology which is seen during autropsies at an incidence of 3/15.000.[1] Systemic venous blood bypasses lungs, and enters into left atrium, and arterial desaturation occurs. Mostly it is a congenital anomaly which consists of multiple fistulas, however it may present as a widely dispersed arteriovenous connections.[2]

Abbreviations:
ECHO Echocardiography
HHT Hereditary hemorrhagic telengiectasis
pAVM Pulmonarey arteriovenous malformation

In this paper a patient with pAVM who applied with complaints of easy fatigability and whose physical examination revealed the presence of cyanosis, and clubbed finger and also underwent transcatheter embolization was presented.

CASE REPRESENTATION

The patient aged eight years and six months had increasingly severe complaints of easy fatigability for the last two years, she had received ambulatory treatment for pneumonia for the last three years, and she was investigated as for the presence of tuberculosis because of a presumed spot on her left lung On physical examination

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clubbed finger was seen (Figure 1a).

Transcutaneous oxygen saturation measured in room air was 75%, and when 100% oxygen was delivered blood oxygen saturation did not increase. His height, and weight percentiles, vital signs, and results of other examinations of body systems were within normal limits. Any abnormal finding was not detected in respiratory sounds, and cardiovascular system examination. Her hemoglobin was 20.4 g/dL, and hematocrit 62%. On chest X-ray a hyperdense area with irregular contours on medial basal region of the left lung was seen (Figure 1b). Her electrocardiographic, and echocardiographic examination findings, and all routine laboratory analyses yielded normal results. She underwent contrast ECHO, and after injection of the contrast material microcavitations were seen on the left atrium, and left ventricle on early phase images, so the test result was considered to be compatible with pAVM. The patient was brought into catheterization suite with the diagnosis of pAVM. Selective contrast material injection into the left pulmonary artery revealed one small, and one large pAVM in the left mid-basal zone. (Figure 2a, b). Occlusion of the large fistula with a diameter of 7 mm was planned using a 14-mm vascular plug-1.

Priorly the large fistula was entered, and control injection with contrast material was performed to be sure of the location to be plugged. Vascular plug 1 was opened in the fistula, and the device was left there. Then 5 mm x 5 cm Gianturco coil was used to occlude the smaller fistula. (Figure 2c, d). Transcutaneous oxygen saturation was 75% before the procedure, but raised to 96% after the procedure. Any postprocedural complication was not seen, and the patient was discharged one day after the procedure At control visits performed within 10 months after the procedure, any problem was not encountered. The family members of the patient gave their informed consent for the use of clinical findings, laboratory test results, and relevant photos.

DISCUSSION

Pulmonary arteriovenous malformation may become manifest in association with congenital abnormalities, chest trauma, Glenn operation, amyloidosis, cystic fibrosis, metastatic carcinomas, and various infections.

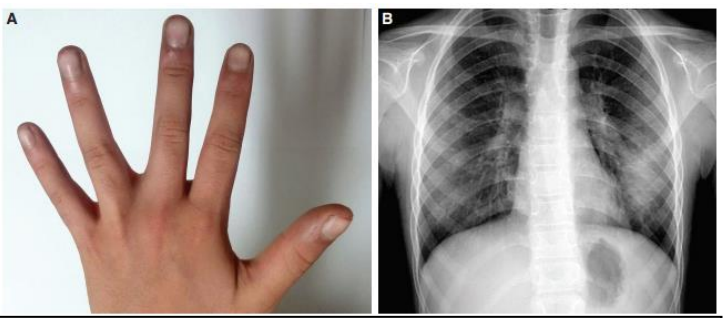

Figure 1. (A) Clubbed fingerr of the patient **(B)** On chest X- ray a hyperdense area in medial basal segment of the lung is seen

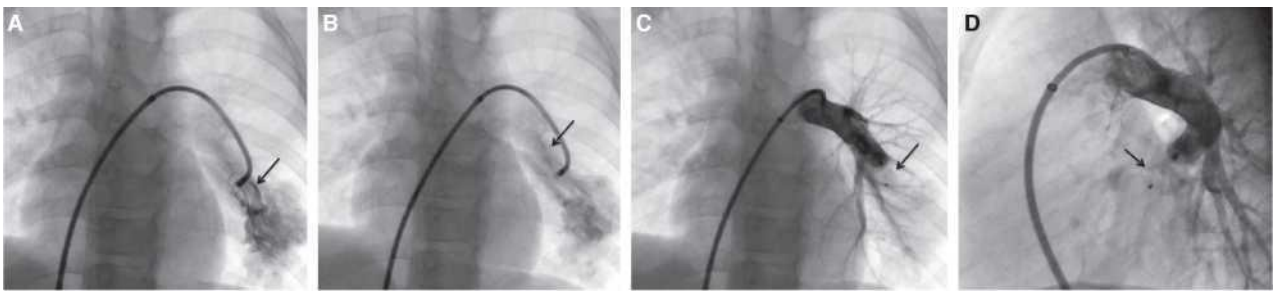

Figure 2. (A, B) Pulmonary arteriogram demonstrates two fistulas between left pulmonary artery, and pulmonary vein. The larger fistula was occluded with Amplatzer vascular plug-I **(C),** and the small one with Gianturco coil (D) Angiogram of the patient after occlusion**.**

Hereditary hemorrhagic telengiectasia with autosomal dominant transmission (HHT disease; Osler-Weber-Rendu syndrome) is frequently associated with widespread pAVM [1] The pathogenesis of the disease has not been fully elucidated yet.

[2] Since in our patient had not relevant family history, epistaxis, telangiectasia or visceral organ involvement, HHT disease was not conceived. Any etiologic factor which might lead to pulmonary arteriovenous malformation was not detected in our patient, so the diagnosis of the patient was presumed to be congenital pAVM.

The patients may be asymptomatic, and symptoms may appear with enlargement of the fistula. If shunt is considerably large then breathing difficulties, cyanosis, clubbed finger, chest pain, nosebleed, hemoptysis, brain abscess related to paradoxical embolism, and stroke may be observed. If the disease left untreated, it may lead to heart failure or infective endocarditis, and if aneurysmal fistula ruptures, then the patient may die.[3] Chest X-ray may demonstrate an opacity if the fistula is large. At the time of diagnosis, contrast ECHO is an important diagnostic tool. Three –eight heart beats after injection of agitated serum through a peripheral vein, observation of bubbles inside the left heart cavities is considered to be a significant finding Computed tomography angiography or conventional pulmonary angiography should be perform in suspect cases. Our patient had consulted pediatricians repeatedly for the complaints of easy fatigability, and coughing, and received treatment for pneumonia at different time periods, and examined as for the presence of tuberculosis. However it was learnt that her transcutaneous oxygen saturation had not been tested before, and during physical examination her clubbed finger, and central cyanosis had not been detected. Apart from easy fatigability, and intermittent complaints of coughing the patient did not have any complaint, and physical examination performed could not reveal any pathologic finding excepting clubbed finger. Her chest X-ray suggested the presence of pAVM, and diagnosis was established based on contrast ECHO findings. Though a consensus does not exist about treatment of pulmonary arteriovenous malformations, but treatment of symptomatic fistulas larger than 3 mm has been strongly advised.[1]

Treatment alternatives are surgical approach, and transcatheter embolization. Surgical approach is appropriate for centrally located single, and large lesions.[4] nIn recent years transcatheter embolization is also frequently applied treatment alternative. This procedure preserves lung parenchyma, and complications related to general anesthesia, and thoracotomy are avoided.

However after transcatheter embolization, complications such as bleeding, thrombosis, displacement of the device may develop, so the patients should be followed up for the occurrence of recanalization.[S6] After embolization procedure oxygen saturation of our patient raised from 75% to 95%, and any complication did not debvelop, recanalization was not observed.

In conclusion, in patients with hypoxemia, and cyanosis, after excluding pulmonary, cardiac, and nervous system pathologies as a rare cause, pAVM should be considered. In some of these patients despite normal physical examination, chest X-ray, ECHO or even computed tomography findings, the patient should be evaluated with contrast ECHO. When the diagnosis of this rarely seen disease is overlooked, one may encounter potentially fatal complications, extreme care should be executed. Transcatheter embolization procedure can be used as a successful treatment modality.

Conflict of interest: None declared.

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Anahtar sözcükler: Çocuk; fistül embolizasyonu; pulmoner arteriyovenöz malformasyon; siyanoz.

Keywords: Child; fistula embolization; pulmonary arteriovenous malformation; cyanosis.