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Case Report / Olgu Sunumu

A two-staged successful transcatheter embolization for multiple pulmonary arteriovenous fistulas

Multipl pulmoner arteriyovenöz fistülün iki aşamalı başarılı transkateter embolizasyonu

Ender Ödemiş, Erkut Öztürk, Hasan Tahsin Tola, Alper Güzeltaş

Department of Pediatric Cardiology, İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

Pulmonary arteriovenous fistulas (PAVF) are abnormal connections between pulmonary arteries and veins through aneurysmal sac or dilated tortuous channels without capillary bed. The clinical presentation includes exercise intolerance and hypoxemia in childhood. Nowadays, transcatheter closure methods using coils, balloons and several devices have become widely accepted treatment modalities, rather than surgical intervention, in childhood. The Amplatzer® vascular plug is frequently used in the management of various pathologies of both peripheral and central vascular structures. In this article, we report a three-year-old boy who underwent a successful two-stage closure of multiple PAVF using an Amplatzer vascular plug (I and II) device.

Key words: Amplatzer vascular plug; child; pulmonary arteriovenous malformation.

Pulmoner arteriovenöz fistüller (PAVF), pulmoner kapiller yatak olmaksızın anevrizmal kese veya dilate kıvrımlı kanallarla pulmoner arterler ve venler arasındaki anormal bağlantılardır. Klinik tablosu çocukluk çağında egzersiz intoleransı ve hipoksemi ile kendini gösterir. Günümüzde çocukluk çağında coil, balon ve çeşitli cihazlar ile transkateter kapatma yöntemleri, cerrahi girişime kıyasla, tercih edilen tedavi seçenekleridir. Amplatzer® vasküler plug, periferik ve santral vasküler yapılara ait çeşitli patolojilerin tedavisinde sıklıkla kullanılmaktadır. Bu yazıda, multipl PAVF'nin Amplatzer vasküler plug (I ve II) cihazı kullanılarak iki aşamalı olarak başarılı bir şekilde kapatıldığı üç yaşında erkek bir olgu sunuldu.

Anahtar sözcükler: Amplatzer vasküler plug; çocuk; pulmoner arteriyovenöz malformasyon.

Pulmonary arteriovenous fistulas (PAVFs) are abnormal communications between the pulmonary arteries and veins without capillary bridging. Arteriovenous fistulas and aneurysmal or cavernous hemangiomas can be characterized as arteriovenous malformations. They are congenital and accompany Osler-Weber-Rendu disease (OWR), also known as hemorrhagic telangiectasia, in the majority of cases.[1,2] In addition, PAVFs cause signs and symptoms like hypoxemia, epistaxis, and dyspnea, and they are part of the differential diagnosis of cyanosis in infants that require treatment. Transcatheter occlusion methods with devices such as coils, balloons, or vascular plugs have now surpassed surgical methods as the most common form of treatment for PAVFs. For example, the Amplatzer™ vascular plug (AVP) (St. Jude Medical Inc., Cardiovascular and Ablation Technologies, Plymouth, MN, USA) has been spectacularly successful at rapidly achieving occlusion.[3,4] This report presents the case of a giant aneurysmal PAVF which was successfully embolized using the Amplatzer® Vascular Plug 1 (AVP 1) and 2.

CASE REPORT

A three-year-old boy was admitted to our clinic for intermittent cyanosis. His personal history indicated that this condition had been episodically present since birth. Physical and laboratory examinations

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Correspondence: Erkut Öztürk, M.D. İstanbul Mehmet Akif Ersoy, Göğüs Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hastanesi, 34303 Küçükçekmece, İstanbul, Turkey.
Tel: +90 212 - 692 20 00 e-mail: erkut_ozturk@yahoo.com

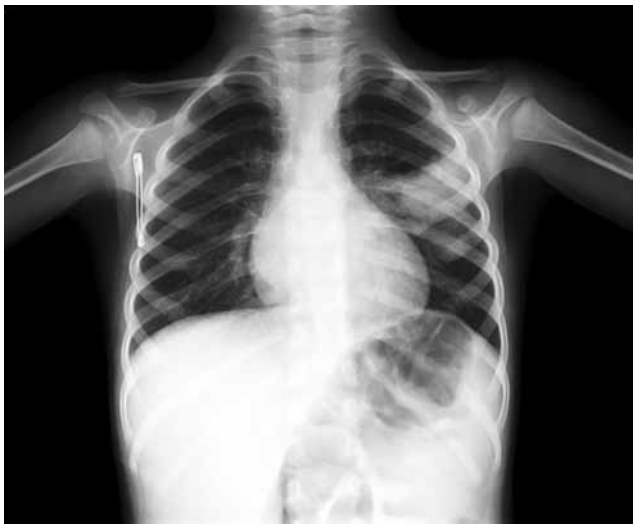

Figure 1. A hyperdense central area as seen on the anteroposterior chest X-ray.

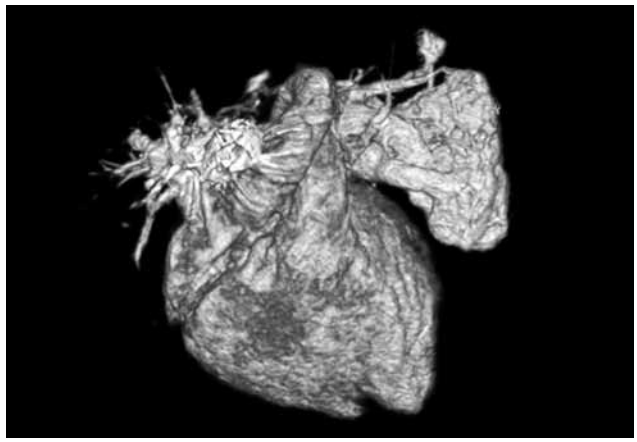

Figure 2. Three-dimensional computed tomography of the chest detected the presence of an arteriovenous malformation.

showed an arterial oxygen saturation of 55%, a heart rate of 120 beats per minute, blood pressure of 120/80 mmHg, and clubbing. A hyperdense central area was seen on the anteroposterior chest X-ray (Figure 1), but the electrocardiogram and transthoracic echocardiography (TTE) were normal. Contrast echocardiography indicated that the left chambers filled up in a shorter time than needed for three cycles. Three-dimensional (3D) computed tomography (CT) of the chest determined the presence of an arteriovenous malformation arising from the branches of the left pulmonary artery that was being fed by three arteries and draining through the left upper lobe pulmonary veins (Figure 2). Pulmonary angiography verified the presence of this malformation being fed by three afferent arteries (Figure 3a, b) with diameters of 9.6, 4.1, and 6 mm, respectively. The 6 mm vessel was plugged by a 12 mm AVP 2 while the smallest vessel was plugged by an 8 mm AVP. Follow-up angiography showed the absence of circulation in the obstructed fistulas and an increase in the blood flow to the areas of the left lung outside the fistula. Due to a prolonged procedure time, an increased use of intravenous contrast material, and a satisfactory pulse oxygen saturation (OS) rate at the end of the procedure, the

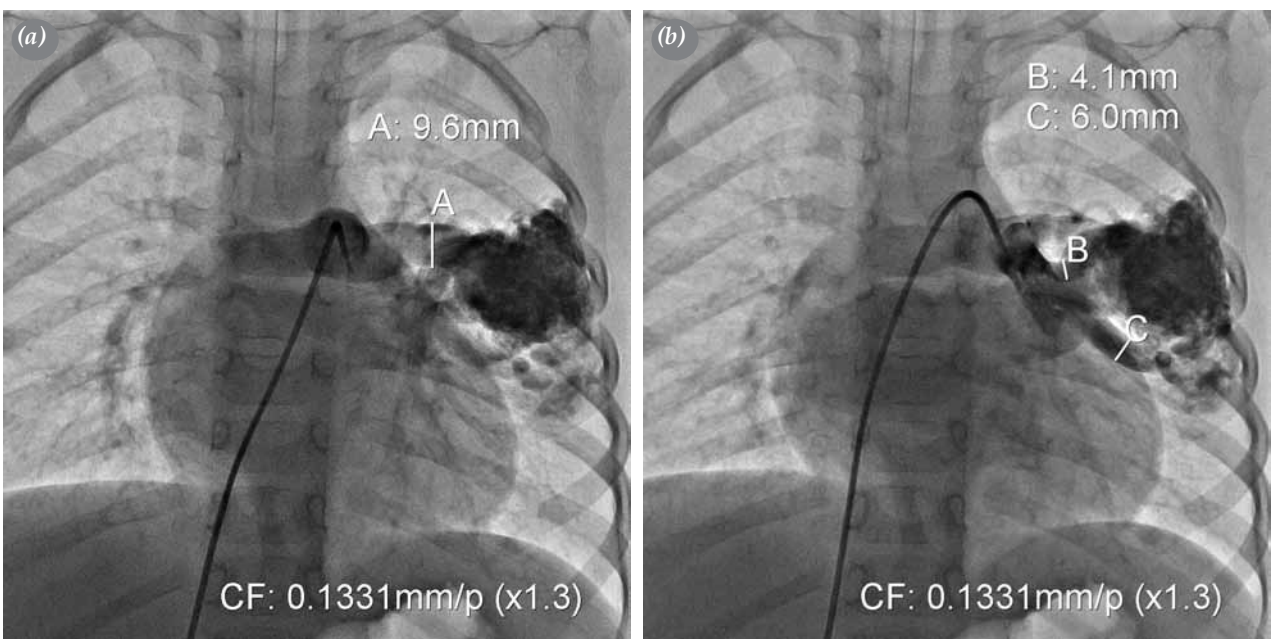

Figure 3. Selective pulmonary angiography showing **(a)** the first feeding artery along with **(b)** the second and third feeding arteries.

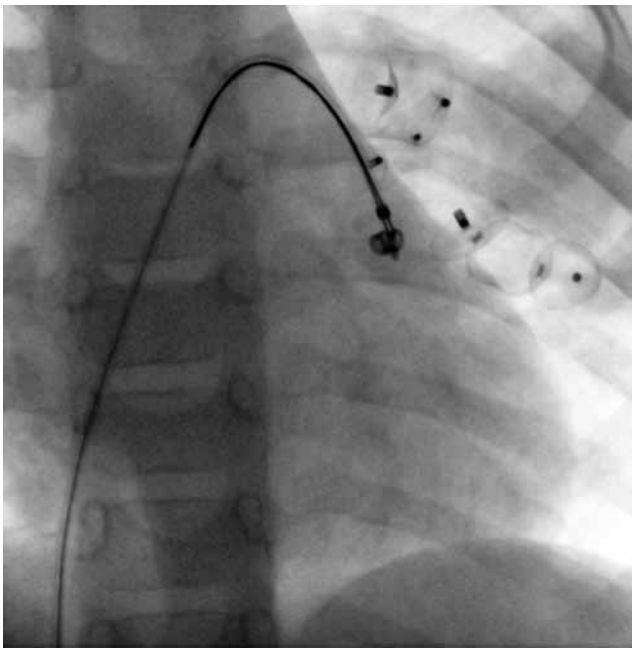

Figure 4. Selective pulmonary angiography after deployment of the vascular plug.

third feeder artery was closed. The intervention was terminated upon achieving an arterial OS rate of 95%. Follow-up measurements one month later showed that the OS rate had fallen to 70%. The third feeder artery (9.6 mm in diameter) was then plugged using an AVP I (Figure 4). The arterial OS breathing ambient air rate was 100% on the day after this intervention, and no loss of OS was observed at the two-month follow-up.

DISCUSSION

Pulmonary arteriovenous fistulas are anomalous direct communications between the pulmonary arteries and veins that bypass the capillary system. Such abnormalities can be either isolated or multiple and may be congenital or acquired. In addition to isolated PAVFs, cases can include those with a positive family history, such as autosomal dominant OWR in which patients can be symptomatic during childhood.[5,6] However, the majority of children with PAVFs are asymptomatic. When symptoms and signs manifest, they can be the direct result of the right-to-left shunt (i.e., asthenia, dyspnea, cyanosis, tachypnea, and tachycardia). Additional manifestations can include hemoptysis, hemothorax, heart failure, paradoxical embolism, and various central nervous system (CNS) complications such as abscess, ischemic attack, seizure, or cerebral hemorrhage.

The diagnosis of PAVFs is usually established via contrast echocardiography, CT, and angiography. Clinical symptoms, for example cyanosis, may recur due to the redistribution of blood in the pulmonary vascular bed after the closure of PAVFs.[7] In addition, increasing age and pregnancy may be predisposing events when such conditions occur. In the current case, our three-year-old patient was reconsulted for cyanosis. A hyperdense area was seen on the chest X-ray, and the contrast echocardiography showed a premature filling of the left heart chambers. The diagnosis of multiple PAVFs was confirmed by a 3D CT scan and pulmonary angiography; however, no genetic abnormality was noted in this patient.

The first choice of treatment for childhood PAVFs is percutaneous transcatheter embolization via placement of a coil or balloon devices. Surgery is reserved for cases in which this type of intervention is unsuccessful or impossible.[8] Angiographic therapy has become the principal treatment for childhood fistulas in recent years. The drawbacks of the coil are essentially the need to use multiple coils, even for a single vessel, incomplete occlusion, coil reflux, and the high proportion of recanalization.[9] The AVP, which is constructed of nitinol wire mesh, represents a new alternative for PAVF treatment, and it has become increasingly popular in recent years due to its reduced frequency of early and late complications, such as embolic events, thanks to its short occlusion time.

Several published case reports have found the AVP to be safe and efficacious for the treatment of childhood PAVF. Çil et al.[3] reported success with nine applications of the AVP in three PAVF patients. No complications were identified, and successful results were noted. The mean follow-up period for that study was six and a half months. Letourneau-Guillon et al.[10] achieved successful occlusion of 35 feeder arteries using 37 AVPs (a 97% success rate) in 35 PAVF patients between the ages of 11 and 86, and they reported no early complications. However, the recanalization of two vessels (7%) was necessary during the 322-day mean follow-up period.

A cost analysis[3,5] has shown that the AVP is cheaper than the Amplatzer™ Duct Occluder (ADO) or the Gianturco-Grifka vascular occlusion device. In addition, it is also less expensive that the vascular plug coil because of its use of multiple coils. To our knowledge, no literature exists which has compared the economic efficiency of the various interventions for PAVF treatment, but the AVP method seems to rank first thanks to its small introducer system and low cost. The relatively high frequency of post-intervention pleuritis, longer intervention duration, and increased

risk of air embolization are the main drawbacks when coils are utilized.

In our case, in the first-stage closure using the AVP II, the patient's OS rate increased to 95% after closing the two PAVFs, but it dropped to 70% at the one-month follow-up visit. This was explained by the redistribution of the pulmonary blood flow because the third AVF did not increase in size, and no new feeder artery was seen on the control angiogram. The last feeder artery was closed using the AVP I, and no complications were encountered during the 180-day follow-up period.

In conclusion, the use of the AVP I and II for the percutaneous transcatheter occlusion of PAVFs during childhood is dependable and effective.

Declaration of conflicting interests

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