

Pulmonary Right-Upper Lobar Arteriovenous Fistula in An Infant with Progressive Cyanosis

Tamer Baysal, MD, Niyazi Görmüş, MD*, Mustafa Cihat Avunduk, MD**, Bülent Oran, MD
Mehmet Yeniterzi*, MD, Sevim Karaaslan, MD

Pediatric Cardiology Unit. *Department of Cardiovascular Surgery and **Department of Pathology
Faculty of Medicine, Selçuk University, Konya, Turkey

Introduction

Pulmonary arteriovenous malformations are malformations providing an abnormal communication between a pulmonary artery and vein that bypasses the normal pulmonary capillary beds. They may occur as an isolated anomaly or as multiple lesions. If larger veins and arteries are affected or if massive involvement of the pulmonary capillaries takes place, an increase in the size of the vessels may occur resulting in severe haemodynamic alterations. Arterial desaturation of oxygen, cyanosis, clubbing of the fingers, and polycythemia may occur, secondary to the intrapulmonary shunt (1).

During the embryological development of the vascular system, which occurs between the 5th and 10th weeks of intrauterine life, a continuous differentiation of the vascular bed occurs, resulting in the creation of separate arterial and venous channels, interconnected by capillaries. When a mistake or halt occurs in this process of vascular differentiation, vascular malformations will appear at different anatomical sites and with variable morphology, depending on the stage of differentiation (1,2).

The etiology of pulmonary arteriovenous fistulae may be congenital or acquired. The congenital form has two varieties: a) cavernous angioma, usually fed by one or more tortuous and dilated branches of the pulmonary artery and b) capillary telangiectasia, which forms a net of capillaries and is usually associated with hereditary hemorrhagic telangiectasia or Rendu-Osler-Weber syndrome (3).

We report the rare case of a 7 month-old boy who had no association with hereditary hemorrhagic telangiectasia but had been presented with intractable hypoxemia and pulmonary arteriovenous fistulae.

Case Report

A 7 month-old male with no significant medical history was admitted to hospital for progressive cyanosis of recent onset refractory to oxygen therapy. He was referred to our hospital because of abnormal shadow in the upper lobe of the right lung that had been seen on a chest radiograph during recurrent pulmonary infections.

Physical examination did not disclose other abnormalities and no additional clinical symptoms were reported except moderate cyanosis and mild fever. There was a grade II-III/VI systolic murmur at the left sternal border. The chest X-ray showed increased vascular markings and irregular infiltration in the upper region of right lung fields with mild cardiomegaly (Fig. 1). Electrocardiography was normal, echocardiography showed trivial mitral and tricuspid regurgitations, patent foramen ovale and no additional cardiac malformation.

Analysis of the arterial gases at room temperature showed severe hypoxemia, with a saturation of oxygen = 41%, hematocrit = 36.8%; hemoglobin = 12.4 g/dL. Serum methemoglobin level was normal. The saturation of oxygen increased only slightly with inhalation of 100% oxygen, which suggested the presence of a large right-left shunt. He did not have Rendu-Osler-Weber syndrome (hereditary hemorrhagic telangiectasia) and also no family history of this syndrome.

The infiltrated area in the right upper lobe persisted despite treatment with seftriaxone and amikacin for 10 days. Then, bubble contrast echocardiography, chest computerized tomographic scan and magnetic resonance angiography demonstrated the arteriovenous fistula in the upper lobe of the right lung (Fig. 2). The right cardiac catheterization was performed to elucidate the diagnosis.

The angiographic study showed a vascular net with diffuse vascular malformations of several large feeding arteries and multiple large venous drainage in the right upper pulmonary lobes (Fig. 3). Pressures in the cardiac cavities were as follows: right atrium = 4 mmHg (mean); pulmonary artery = 11 mmHg (systolic), 6 mmHg (diastolic), 9 mmHg (mean).

After the right upper lobectomy, the cyanosis completely disappeared. The immediate result was an increase in arterial oxygen saturation from 40% to 95-98%. At the time of hospital discharge, oxygen saturation measured at room temperature with the pulse oximeter was more than 90%. Following a successful surgical procedure, the patient had no serious postoperative complications. The diagnosis was also confirmed histopathologically. The tissue specimen was examined in light microscopy with Hematoxylin-Eosin. An abnormal communication between pulmonary artery and vein has been seen. Several dilated and cavernous capillaries were filled with eryth-

rocytes (Fig. 4). The patient has been well for the following two months.

Discussion

Pulmonary arteriovenous fistula is a rare vascular abnormality in children. It may show few symptoms. Progressive cyanosis refractory to oxygen therapy without any apparent cardiac or pulmonary disease is strongly suggestive of pulmonary arteriovenous fistula. It is not easily diagnosed routinely. One of the simple and easy diagnostic investigations is contrast echocardiography (4), in which agitated saline solution, for example, can be injected in a peripheral vein. The microbubbles resulting from this process detected in the left atrium allow to determine a right-to-left shunt through the lungs, as in our case. The capillary alveolar filter does not allow the passage of these microbubbles. There are other radiodiagnostic methods in this subject as thorax computerized tomography, magnetic resonance angiography and right cardiac catheterization.

Interest in pulmonary arteriovenous malformati-



Figure 1. Chest x-ray shows irregular infiltration in the upper lobe of right lung.

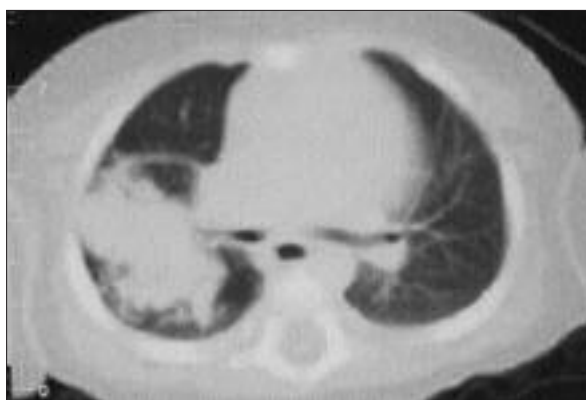


Figure 2. Magnetic resonance angiography demonstrates the arterio-venous fistulas in the upper lobe of the right lung.



Figure 3. Angiographic study shows a vascular net with diffuse vascular malformations in the right upper pulmonary lobe.

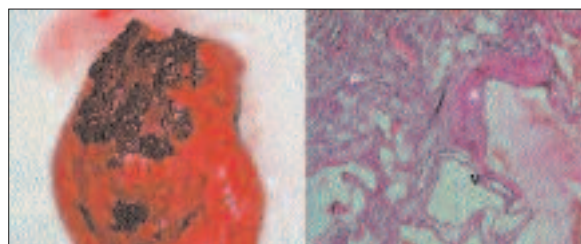


Figure 4. Light microscopy with Hematoxylin-Eosin of the tissue specimen. Several dilated capillaries and an abnormal communication between a pulmonary artery (A) and pulmonary vein (V) in the pulmonary parenchyma (P) are seen.

ons has increased since the mid 1980's with the realization that they cause more clinical problems and a greater morbidity than had been previously recognized, together with the emergence of transcatheter embolization as an effective and safe alternative therapy to surgery (1). For twenty years better interventional catheterization techniques have been developed and our understanding of the clinical outcomes of these lesions has improved. Coil embolization should be considered more often for the treatment of arteriovenous malformations because it is safer, more effective and less invasive than traditional operations (4-8). Another alternative approach would be occlusion of each pulmonary lobe with a detachable balloon, and concomitant assessment of blood saturation, therefore checking whether selective embolization with detachable balloon could replace pneumectomy (9).

It is worth emphasizing that blood saturation may decrease with time as a consequence of opening new fistulae because of the progressive character of the disease (10). In our case, cyanosis was of progressive character and there was no cyanosis during first 3-4 months of the life. His diagnosis was also confirmed histopathologically as diffuse cavernous arteriovenous fistula. We chose the surgical approach, because of huge, cavernous and progressive character of the arteriovenous malformations, bounded by only right upper lobe of the lungs.

Finally, we think that surgical therapy is safer than interventional techniques for progressive and huge arteriovenous malformations, especially in case of unifocal localizations.

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