CASE REPORT

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For the video/videos of the article:

# Amplatzer Vascular Plug Occlusion in a Child with Late-diagnosed Multiple Pulmonary Arteriovenous Malformations

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**ABSTRACT** Pulmonary arteriovenous malformations (PAVMs) are abnormal direct connections between the pulmonary artery and pulmonary vein, which may result in a right to left shunt. Patients may be asymptomatic, but symptoms may gradually occur with the enlargement of the fistula. A 5-year-old Syrian female patient was followed by the primary care physician during the last year with a diagnosis of polycythemia. The patient reported headache and abdominal pain for the last three months and was referred to the pediatric cardiology outpatient clinic when clubbing was detected. Normal sinus rhythm was observed in electrocardiography and echocardiographic examination findings were normal. Thoracic computed tomography/angiography was performed and PAVM was detected in the upper segment of the lower left lobe. Catheter angiography was performed with the diagnosis of PAVM. The fistulas were closed using three vascular plugs. Transcutaneous oxygen saturation increased to 96% after the procedure.

**Keywords:** Pulmonary arteriovenous fistulas; therapeutic embolization

Pulmonary arteriovenous malformations (PAVMs) are abnormal direct connections between the pulmonary artery and pulmonary vein, which may result in a right to left shunt.[1] Patients may be asymptomatic, but symptoms may gradually occur with the enlargement of the fistula. If the amount of shunt is excessive, respiratory distress, cyanosis, clubbing, chest pain, epistaxis, hemoptysis, brain abscess due to paradoxical embolism and stroke may be seen. Heart failure or infective endocarditis may occur in patients without treatment, sudden death caused by rupture of the aneurysmal fistula may even be seen.[1,2] Non-infectious opacity may be seen in chest X-ray. Computed tomography angiography or conventional pulmonary angiography is required in patients with a suspected diagnosis.[3] Transcatheter embolization of abnormal vascular connections is the current treatment method in this disease.[4]

We aimed to present our patient who was diagnosed as having PAVM while being followed for polycythemia and successfully underwent transcatheter fistula embolization.

## CASE REPORT

A 5-year-old Syrian female patient had been followed by the primary care physician for the last year with a diagnosis of polycythemia. The patient reported headache and abdominal pain for the last three months and was referred to the pediatric cardiology

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outpatient clinic when clubbing was detected. The patient had no history of cough, dyspnea, chest pain or palpitations and was active for her age and able to keep up with her peers. Physical examination showed a well-developed, nondysmorphic child. Her height and weight were 104 cm (50th-75th percentile) and 16.7 kg (50th-75th percentile), respectively. The blood pressure was 99/61 mmHg (within the normal range for age), heart rate was 110 beats/minimum, and respiration rate was 28 breaths/minimum. There was a mild bluish discoloration of the oral mucosa. A cardiovascular examination revealed a regular rhythm with a normal S1 and S2. There was no murmur, rub or gallop. Her abdomen was soft and not distended, with no organomegaly or mass. The extremities were normal with no edema. However, there was clubbing in the fingers and toes. The oxygen saturation was 75% in room air. In laboratory analyses, hemoglobin was found to be 18.7 g/dL and the hematocrit was 57%. On chest X-ray, a hyperdense area with irregular contours was seen in the medial basal region of the left lung (Figure 1). Normal sinus rhythm was observed in electrocardiography and echocardiographic examination findings were normal. Thoracic computed tomography/angiography was performed and PAVM was detected in the upper segment at the level of the lower left lobe (Figure 2). Catheter angiography was performed with the diagnosis of PAVM. In the selective contrast injection into the left pulmonary

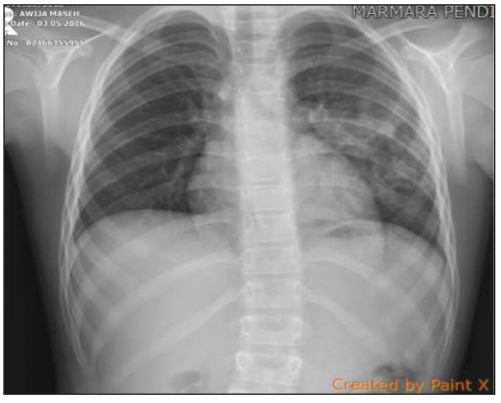


**FIGURE 1:** Chest X-ray.

artery, it was observed that there were three PAVMs in the left middle-lower zone (Figure 3, Video 1). The fistulas were closed using three vascular plugs, 10 mm, 12 mm, and 14 mm in size (Figure 4, Video 2). Transcutaneous oxygen saturation increased to 96% after the procedure. No postprocedural complications were seen, and the patient was discharged on the day after the procedure. Informed consent was obtained from our patient 's family, and they signed permis-sion to publish her story.

## DISCUSSION

PAVM is mainly congenital but may also occur due to thoracic trauma, Glenn's procedure, amyloidosis, cystic fibrosis, metastatic carcinomas, and various infections.[1,2] The coexistence of autosomal dominant

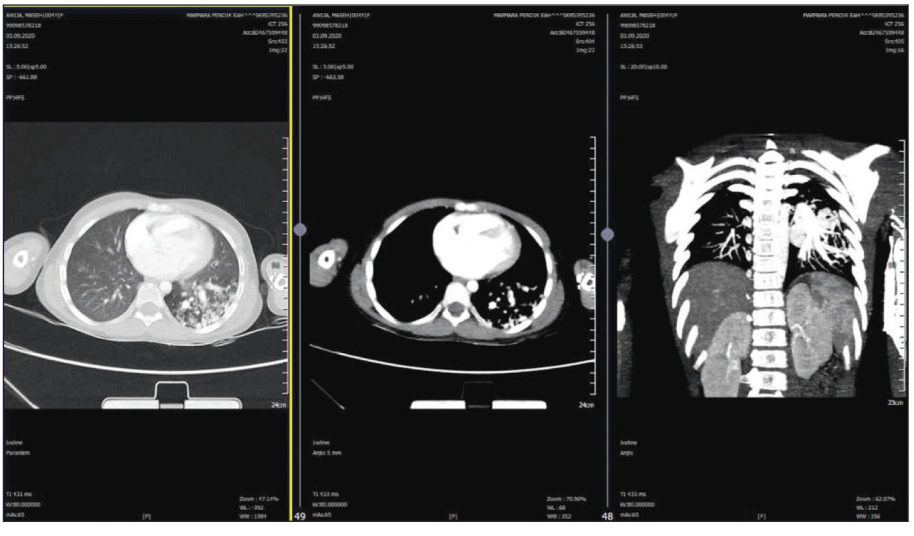

**FIGURE 2:** Computed tomography of the thorax.

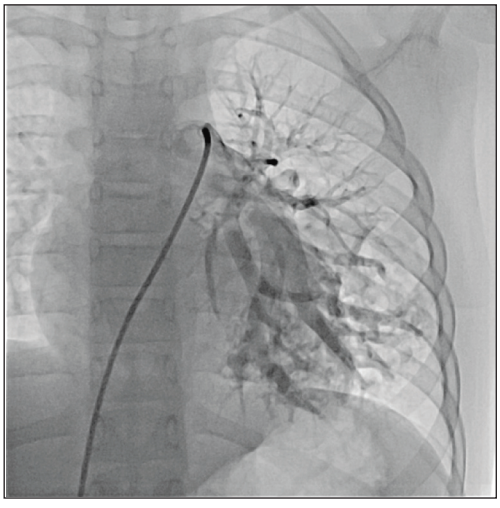

**FIGURE 3:** Angiographic anteroposterior view before the procedure.

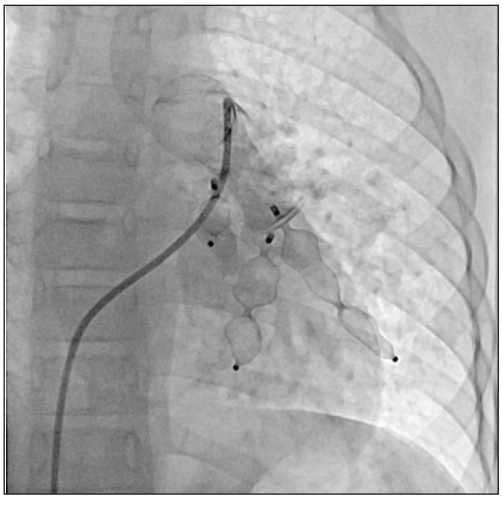

**FIGURE 4:** Angiographic anteroposterior view after the procedure.

hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome) and extensive PAVM is common.[5] The pathogenesis of the disease is not fully understood.[3,4] Hereditary hemorrhagic telangiectasia was not considered in the patient because there was no family history, epistaxis, telangiectasia or visceral organ involvement. No specific cause of PAVM could be determined, so the patient was accepted as having congenital PAVM. Although there is no consensus on the treatment of PAVM, it is stated that fistulas larger than 3 mm and patients with symptoms should be treated. Treatment options are surgery and transcatheter embolization.[4,6] Surgical intervention is suitable for centrally located, single, and large lesions. In recent years, transcatheter embolization has been commonly used because it offers more advantages in terms of lung parenchyma preservation and complication avoidance related to thoracotomy.[4,6] However, complications such as bleeding, thrombus, and device displacement may develop after transcatheter embolization, and patients should be followed up in terms of recanalization after the procedure.[4,6] The oxygen saturation of our patient increased from 75% to 96% after the embolization procedure and no complications or recanalization were observed. As a result, PAVM should be considered as a rare cause of hypoxemia and cyanosis in patients without lung, heart, and nervous system pathologies. Even if a physical examination, chest X-ray, and echocardiography are normal, patients should be evaluated using contrast echocardiography or chest computed tomography angiography.[3] When the diagnosis of this very rare disease is delayed, heart failure may develop.[3,6] In treatment, transcatheter embolization can be successfully performed.

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### *Conflict of Interest*

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

### *Authorship Contributions*

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