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PULMONARY ARTERIOVENOUS MALFORMATION: A RARE CAUSE OF CYANOSIS IN A CHILD

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Abstract

Pulmonary arteriovenous malformations are rare, abnormal low resistance vascular structures that connect a pulmonary artery to a pulmonary vein, thereby bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt.

We report a 11 -year - old boy who presented with fatigue, cyanosis since the age of 10. Computed tomography pulmonary angiogram showed a large pulmonary arteriovenous malformation (PAVMs) at the left and right lung.

Most of the multiple arteriovenous malformations were occluded with Amplatzer vascular plug. He is now still cyanotic, with blood oxygen saturation of 88%.

Keywords: arteriovenous malformation, cyanosis, pulmonary arteriovenous malformation, pulmonary vascular anomaly.

Introduction

PAVMs are rare, abnormal low resistance vascular structures that connect a pulmonary artery to a pulmonary vein, thereby bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt. The first description of pulmonary arteriovenous malformation was reported by Churton in 1897[1].

Symptoms vary from easy fatiguability and cyanosis because of intrapulmonary shunting. The main complications of PAVMs result from intrapulmonary shunt and include stroke, brain abscess and hypoxemia.

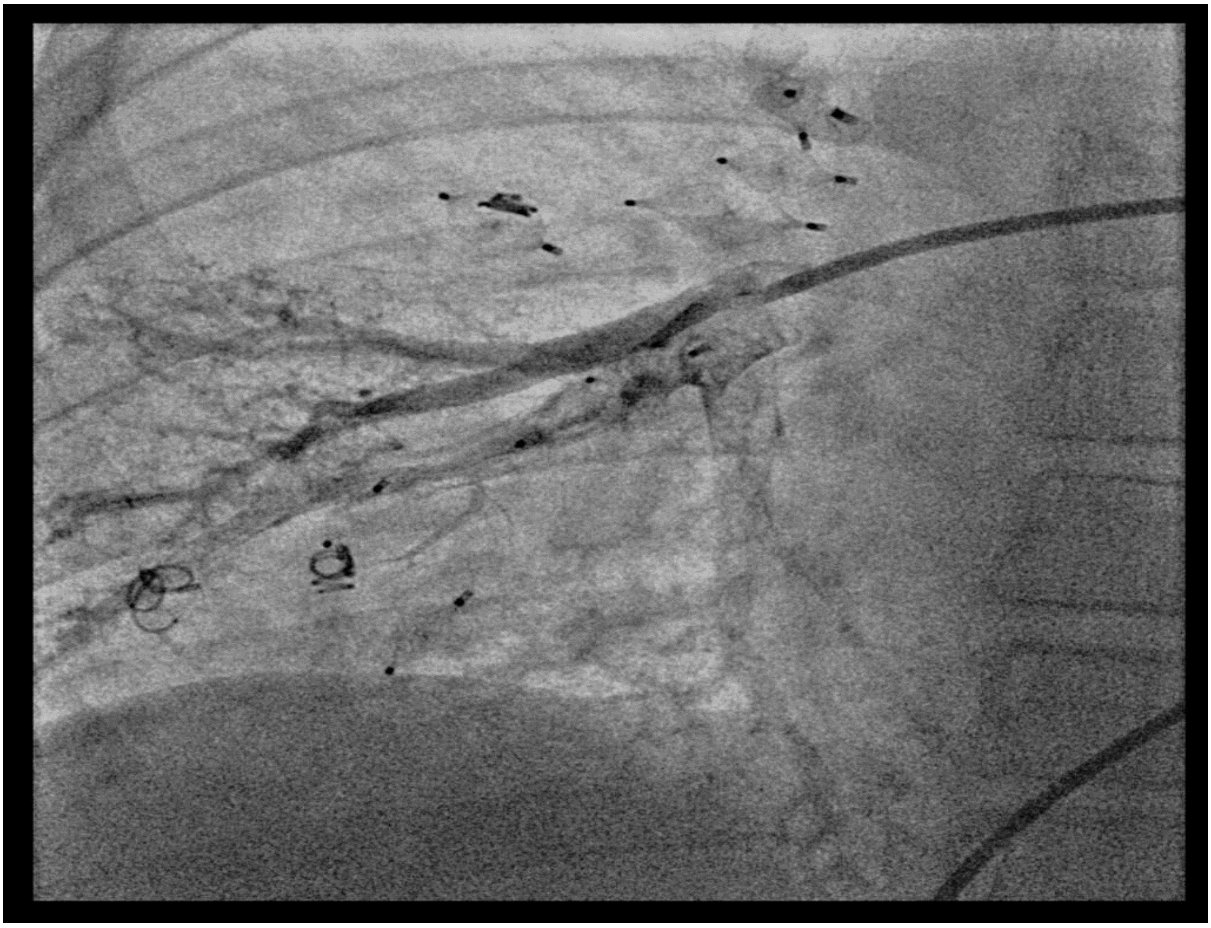
Local pulmonary complications include PAVM rupture leading to life-threatening hemoptysis or hemothorax. The preferred treatment of choice is transcatheter occlusion of the feeding artery.

CASE REPORT

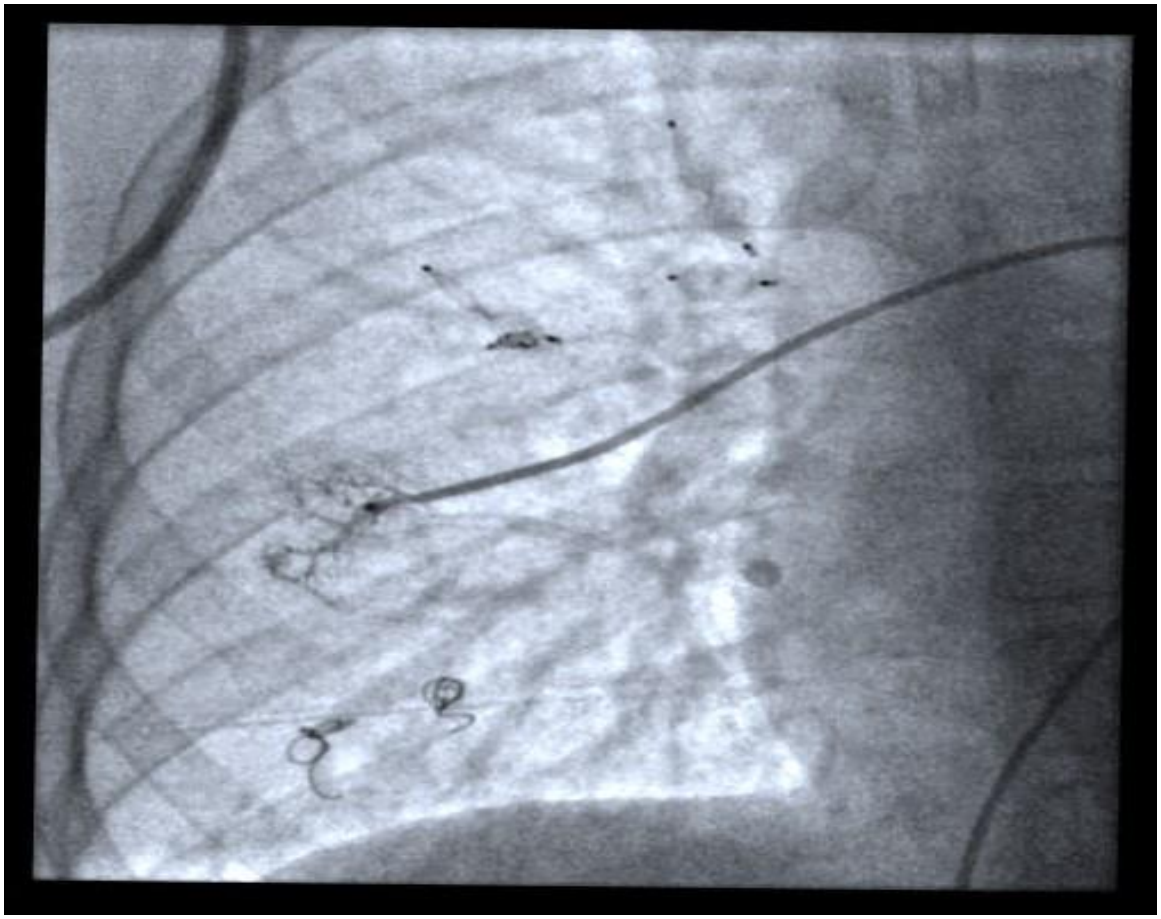
An 11 -year -old boy presented with fatigue, decreased exercise tolerance and cyanosis occurring 10 months before the diagnosis. There was no chest pain, palpitations, neurological symptoms, atopy, hemorrhagic tendency or history of chest trauma.

On the skin on the hands and legs, 5 vascular stigmata-hemangiomas were seen. He was cyanotic with pulse oximetry of 75% on room air. Complete blood count revealed polycythemia (Er 6.3x10^12, Hb 189g/l, Hct 55.2%). Chest radiograph showed a homogeneous soft tissue mass of the both lungs. Transthoracic echocardiography showed normal cardiac structures with no evidence of pulmonary hypertension.

Computed tomography (CT) pulmonary angiogram showed a large pulmonary arteriovenous malformation of the right and left lung. Transcatheter pulmonary angiogram was perform twice and showed a large arteriovenous malformation in the both lungs with a feeding vessel bigger of 2 mm and few small left lung arteriovenous malformations with feeding vessels of 2 mm each (Figures 1 and 2).



Figures1 and 2. Percutaneous transcatheter embolization on the right side



Figures1 and 2. Percutaneous transcatheter embolization on the right side

Most of the arteriovenous malformations was occluded with Amplatzer vascular plug (Figure 3).

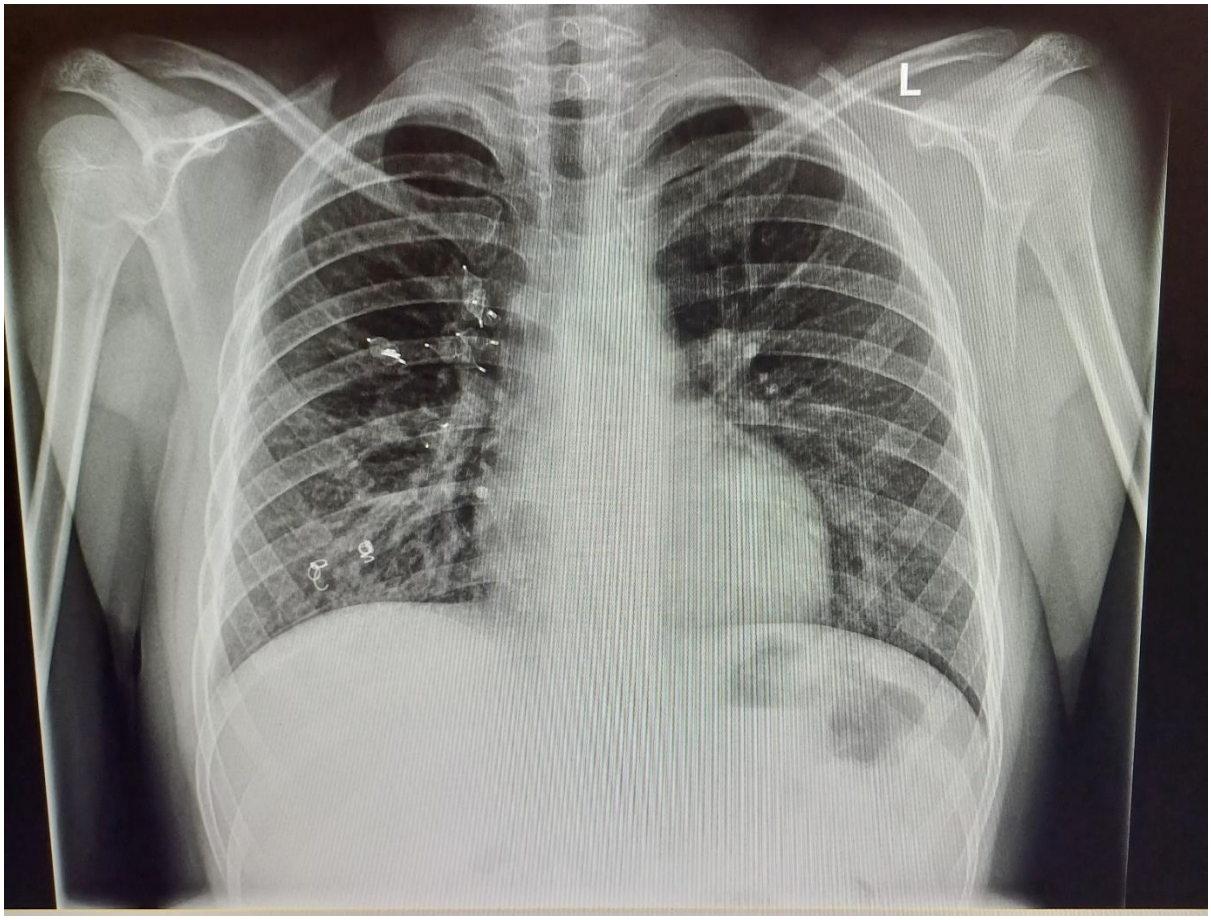


Figure 3. Chest X-ray showing multiple embolization coils on the right side

The percutaneous pulse oximetry saturation had risen from 75% to 88% immediately, on room air. Currently, his exercise tolerance has improved, but he is still cyanotic. His follow-up was one month post transcatheter occlusion, then six -monthly. He will need further follow up for another catheterization and occlusion of the remaining AV-malformations.

Discussion

Pulmonary arteriovenous malformations are rare, occurring in 2-3 per 100,000 population. 80% of them are congenital, and rarely due to secondary conditions such as post congenital heart disease surgery as modified Fontan procedure, Glenn procedure or hepatopulmonary syndrome. 47%-80% are associated with hereditary haemorrhagic telangiectasia [2].

The afferent supply can include the pulmoary artery, aorta, intercostals and bronchial arteries. The efferent limb drains into the pulmonary vein, left atrium or inferior vena cava. Chest radiography detects 98% of arteriovenous malformations, and contrast - enhanced CT thorax detects 90% of arteriovenous malformations [3].

Typical PAVMs, on chest radiography, should appear as well-defined lesions with feeding vessels. This imaging modality, has a low sensitivity for detecting smaller sized PAVMs. However, pulmonary angiography is still the gold standard.

There is evidence that pulmonary arteriovenous malformations progressively enlarge over a period of time. The main complications of PAVM result from intrapulmonary shunt and include stroke, brain abscess, and hypoxemia.

Local pulmonary complications include PAVM rupture leading to life-threatening hemoptysis or hemothorax. The preferred treatment of choice is transcatheter occlusion of the feeding artery [4,5,6].

However complete cure is rare and collateralization of closed PAVMs and/or progressive enlargement of the small ones following treatment may occur, and hence long- term clinical and imaging follow-up is required to assess PAVM enlargement and PAVM reperfusion.

They may need to be repeated catheterizations during follow- up for further need of transcatheter occlusion of newer enlarged PAVMs.

Complications of transcatheter occlusion are pleuritic chest pain, pulmonary infarction, myocardial rupture, stroke, deep venous thrombosis, vascular injury, device migration and early balloon deflation [6].

Follow-up screening is indicated at one-month after catheterization and yearly thereafter. Spiral CT thorax should be done every 3-5 years to look for development of new or growth of small pulmonary arteriovenous malformations [3].

Antibiotic prophylaxis is recommended for any procedure that may induce bacteremia. Surgery is indicated for failure of transcatheter occlusion, serious bleeding, and intrapleural rupture of the pulmonary arteriovenous malformation[5].

The techniques are local excision, segmental resection, lobectomy, ligation and pneumonectomy.

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