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Symptomatic Pulmonary Arteriovenous Malformations in a 10-Year-Old Boy – A Case Report

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Abstract

Introduction: Pulmonary arteriovenous malformation (PAVM) is a rare cardiovascular anomaly and represents direct communications between the branches of pulmonary artery and pulmonary veins, without an intervening pulmonary bed.

The incidence of PAVM is 2–3 per 100,000 population. The male to female ratio varies from 1:1.5 to 1.8. A clinical manifestation may be from asymptomatic to severe hypoxia.

Case presentation: A ten years old boy was admitted to a regional hospital because the mother noticed that the boy was slower in the game at the teacher noticed that the boy was slower and could not lift his eyes up of clinical and laboratory examinations have been shown polycythemia, and cyanosis, bat-like appearance of the fingers and polycythemia, RTG chest and CT have claimed AV malformation of the lower left lobes with the feeding artery Neurologic symptoms may be the presenting symptoms in up to 40% of patients.

The patient was successfully treated with left lower lobectomy. Early post-operative recovery has been successful. Later checkups showed better blood saturation with oxygen, the same polycythemia, and a better clinical condition.

Discussion and Conclusion: If the etiology of the hypoxemia often remains unclear pulmonary AVM should be considered. The diagnosis needs to be confirmed with a CT scan that should identify the food artery.

If the AV malformation occupies large part of the lobe, a reasonable treatment options should be lobectomy. Long-term follow up, including chest CT examinations every 1 to 2 years, is recommended.

Keywords: Arteriovenous malformations; Lung; Hypoxia

Introduction

Pulmonary arteriovenous malformation (PAVM) is a rare cardiovascular anomaly. PAVM represent abnormal communications between the pulmonary arterial and venous systems that bypass the capillary bed.

The incidence of PAVM is 2–3 per 100 000 population. The male to female ratio varies from 1:1.5 to 1.8. A clinical manifestation may be from asymptomatic to severe hypoxia [1].

Pulmonary PAVM may be classified as simple with a single feeding and draining vessel (80% of cases), or complex, with 2 or more feeding or draining vessels (20% of cases). Up to 65% of pulmonary AVMs are found in the lower lobes of the lung [2].

Case Presentation

A 10-year-old boy was admitted to a regional hospital because of the following symptoms: the mother noticed that the boy was slower in the game; the teacher noticed that the boy was slower and could not lift his eyes up.

After that, the child is transferred to the Cardiology of the Pediatric Clinic. Physical examination reveals mild cyanotic coloration of peripheral parts and bat-like appearance of the fingers **(Figure 1).**

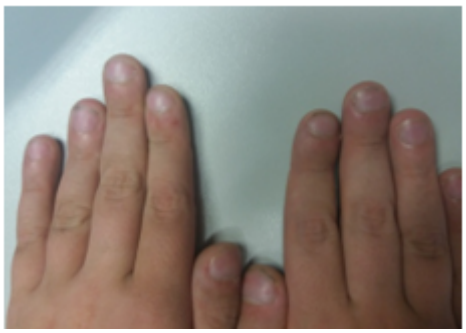

Figure 1 Bat-like appearance of the fingers.

Laboratory findings show eritorcitosis (Er=6.28, Hgb=173, Hct=0.52) and saturation of the oxygen 73%. The chest RTG shows the shadow on the left side. CT chest that shows the existence of a large AV malformation in the area of the lower left lung (**Figure 2**). Which is feeding by artery from the lower left lung artery, and the drainage vein is drained in the lower left pulmonary vein (**Figure 3**).

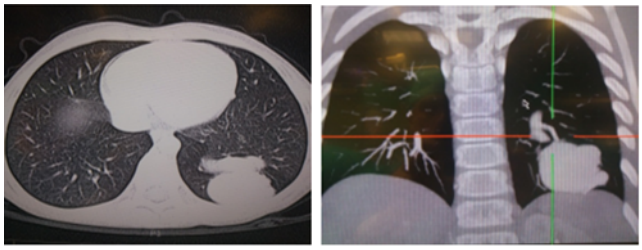

Figure 2 CT Chest that shows the existence of a large AV malformation in the area of the lower left lungs.

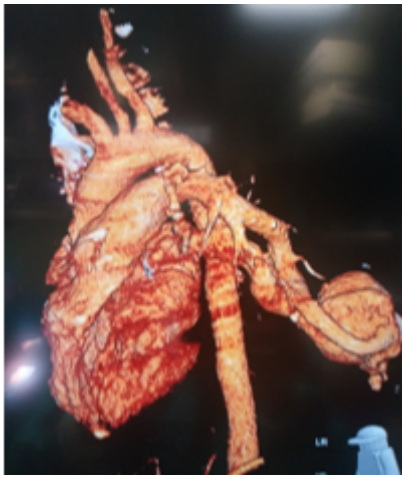

Figure 3 CT feeding artery from the lower left lung artery and the drainage vein is drained in the lower left pulmonary vein.

Other reviews

ECHO hearts, a 24-hour ECG Holter monitoring, EEG, TCD excluded the possibility of other telangiectasia, and spirometry was performed to exclude obstructive disorders.

The patient was surgically treated, left thoracotomy was performed. In the left lower lobe of the lung was verified flank mass, which had a diameter about 1 cm **(Figure 4).**

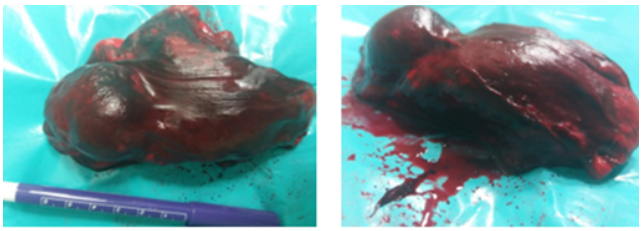

Figure 4 Left lower lobe of the lungs with associated prominent AV malformation.

Typical left lower lobectomies with associated AV malformation were performed. Early postoperative treatment was satisfactory. The patient recovered. Chest RTG was satisfactory **(Figure 5).**

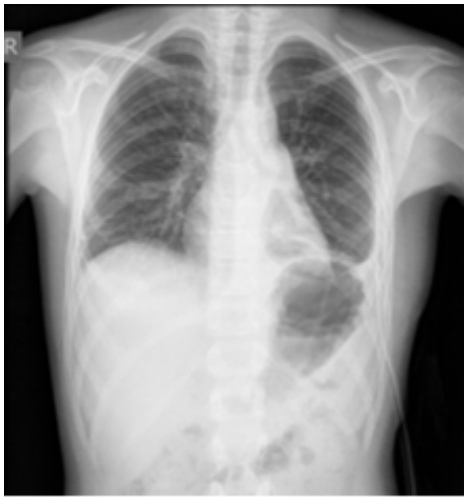

Figure 5 Post-operative RTG chest.

Microscopic examination showed multiple anastomosing dilated vascular channels with irregularly thickened, abnormally distributed through the lung **(Figures 6-8).**

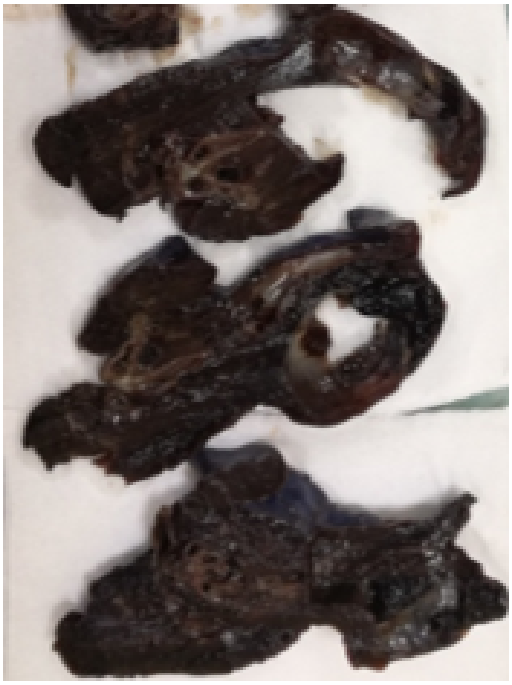

Figure 6 Arteriovenous malformation. Grossly, multiple ectatic vessels in lung parenchyma.

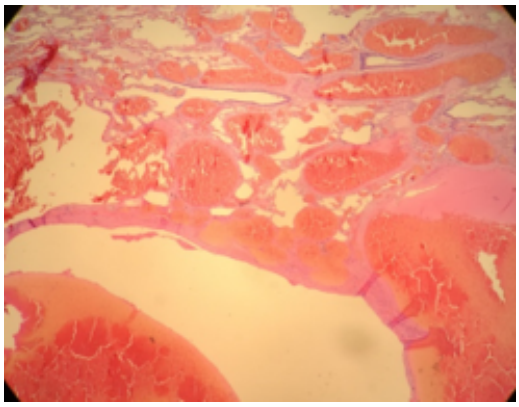

Figure 7 Microscopic examination showed multiple anastomosing dilated vascular channels with irregularly thickened, abnormally distributed through the lung (Hematoxylin-eosin staining x10).

He was discharged from the hospital on the 10th day of hospitalization. Control examination 3 months after surgery showed a regular respiratory status, neat saturation with oxygen 95% in comparison with preoperative which was 73%, less difficulty in physical activity, identical polycythemia as preoperative, and somewhat less pronounced “bat-like” appearance of the fingers. Parents were pleased with improving their child's physical abilities. Long-term follow up, including chest CT examinations for 1 year, was recommended.

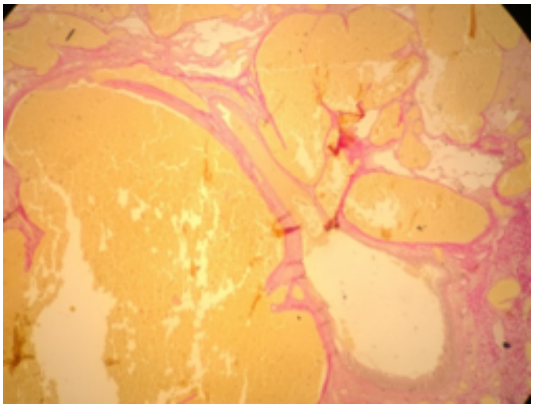

Figure 8 Vessels walls are thickened by fibrous tissue and shows deficiency of smooth muscle making difficult to differentiate veins and arteries (Elastic van Gieson staining x10).

Discussion

Systemic hypoxemia in children’s is commonly due to pulmonary parenchymal disease, pulmonary hypertension with atrial level right-to-left shunting, structural heart disease, or a severe neurological insult. If the etiology remains unclear, pulmonary AVM should be considered. From 50% to 90% of pulmonary AVM are associated with hereditary hemorrhagic telangiectasia (HHT), Rendu-Osler-Weber syndrome [3]. Our patient was like most patients which had solitary PAVM seen in the left lower lobe being as the most common location. The majority of multiple PAVMs are also confined to bilateral lower lobes; the incidence of bilateral PAVMs ranges from 8% to 20% [4].

Pulmonary AVM frequently go unrecognized until the late teens, but they may remain asymptomatic throughout life. Clinical presentation of PAVMs ranges from an incidental finding on a chest roentgenogram in an asymptomatic patient to polycythemia, cyanosis, and congestive heart failure. Neurologic symptoms may be the presenting symptoms in upto 40% of patients [5].

The first patient with AV malformation was described during a pathological study one century ago [6]. The options for treating this malformation throughout history were different. The first reported surgical intervention for PAVM was a pneumonectomy in 1940 by Shestone for a large centrally located lesion [7]. The first transcatheter occlusion of a pulmonary AVM was described in 1978 with metal coils, and 1993 with balloon occlusion [8].

We report our experience of 10-year-old boy with refractory hypoxemia due to pulmonary AVM. We emphasize a strong suspicion for the pulmonary AVM in infants with systemic hypoxemia, cyanosis, “bat-like” appearance of the fingers, slow psychomotor development, pronounced polycythemia of unclear etiology and a structurally normal heart.

Additional diagnostic tests are chest radiography and CT, could find a pulmonary AVMs as rounded, circumscribed pulmonary nodules. Feeding vessels could be visualized. On chest CT, a homogeneous, circumscribed, non-calcified nodule may be seen. Likewise, a serpiginous mass may be seen with connections to vascular structures.

Pulmonary AVM treatment is recommended for symptomatic patients or those AVMs with a feeding artery diameter 3 mm [9]. Transcatheter embolotherapy with stainless steel coils or detachable balloons is most commonly performed [10]. Following embolotherapy, a considerable decrease in pulmonary AVM size is expected; persistent size may indicate persistent perfusion.

Conclusion

Conservative surgical resection remains the treatment of choice. Due to the size of AV malormation which occupied a large part of the left lower lobe we decided to perform the lower left lobectomy. Long-term follow up, including chest CT examinations every 1 to 2 years, was recommended. If the etiology of the hypoxemia often remains unclear pulmonary AVM should be considered. The diagnosis needs to be confirmed with a CT scan that should identify the feeding artery. If the AV malformation occupies large part of the lobe, a reasonable treatment options should be lobectomy. Long-term follow up, including chest CT examinations every 1 to 2 years, is recommended.

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