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Pulmonary Arteriovenous Malformations Incidentalloma in a 10-Years-Old Child

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Abstract

Pulmonary Arteriovenous Malformations (PAVMs) are structurally abnormal vascular communications between pulmonary arteries and pulmonary veins, which bypass the normal capillary bed and cause a low resistance right-to-left shunt with refractory hypoxemia. Generally, PAVMs were congenital, most commonly associated with (Hereditary hemorrhagic teleangiectasia (HHT). The age of diagnosis is very variable, range neonatal to adulthood, mostly diagnosed in the first 3 decades of life and clinical manifestations occur later in life generally. Here, we report PAVMs discovered incidentally in a 10-years-old child without any known risk factor.

Keywords

pulmonary arteriovenous malformation, communications between pulmonary arteries and pulmonary veins, pediatric

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Introduction

Pulmonary arteriovenous Malformations (PAVM) are generally congenital lesions corresponding to abnormal communications between the pulmonary arterial circulation and the pulmonary circulation.[1,2] They can be isolated form or most commonly in association with hereditary hemorrhagic telengiectasia (HHT), also kwon as Osler-Weber-Rendu syndrome.[1] Symptoms depending on the size and number of PAVMs become more evident in adults. They are not commonly diagnosed in pediatric age.[2] Here, we report isolated form PAVMs discovered accidentally in a 10-years child.

Case Report

Ten-years-old children, without pathological history, come to the emergency for inhaling foreign object (whistle ball). On admission he was a good general condition, conscious, a little anxious, no cyanosis, respiratory rate 17/minute, heart rate 90/minute, blood pressure = 120/70 mmHg, oxygen saturation was 85%. There were no signs of right heart failure or pulmonary hypertension. He was put on oxygen therapy 15l/minute, oxygen saturation ranging from 85% to 87%. Arterial blood gas revealed pH: 7.46, pCO_2:35 mmHg, pO_2:54 mmHg. Others biological examination was normal. He was transferred to intensive care to extract the whistle ball and was removed without incident. The ball of the whistle was at the level of the right main bronchus just after the bifurcation. After, the child was stable but the oxygen saturation was between 85% and 87% with oxygen therapy. Arterial blood gas control was pH: 7.44, pCO_2:34 mmHg, pO_2:55 mmHg. Others biological examination was normal.

Chest radiography showed homogenous area of increased density with regular border at the right paracardiac without cardiomegaly, (Figure 1). Chest X-ray was performed after extraction of the foreign object. No previous chest X-ray was available.

We completed by chest computerized tomography (CT*) without contrast showed a large soft tissue oval, proximal as vascular nature in the posterior-segment of the right upper lob. CT scan with injection confirmed the diagnosis of a simple large serpiginous right pulmonary arteriovenous fistula. The right pulmonary

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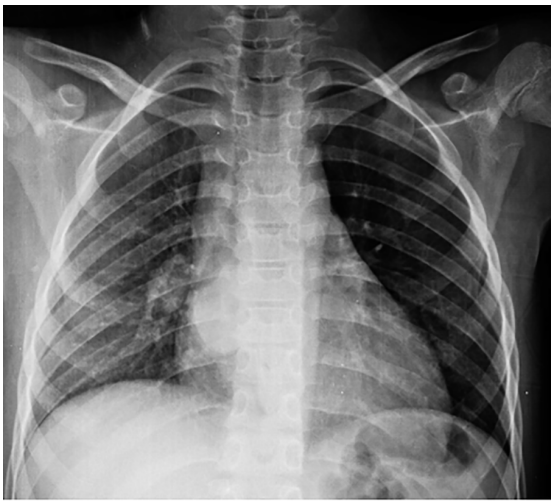

Figure 1. Chest X-ray: homogenous area of increased density with regular border at the right para-cardiac without cardiomegaly.

artery communicating with the right inferior pulmonary vein (Figures 2-4).

Echocardiography was performed and was normal but arteriovenous malformation has not been viewed.

The evolution was marked by clinical stability of the respiratory system with an oxygen saturation not exceeding 87% under oxygen therapy and 85% in ambient air. After careful investigation, we found no familial risk factors for Osler-Weber-Rendu syndrome or HHT objectively. However, the child reported that at school, during physical exertion, he tired faster than his peers, but no previous hospitalization was reported. Thereby, we considered that is a probable congenital PAVM. The child was referred to the service of pediatric surgery.

Discussion

PAVMs are structurally abnormal vascular communications between pulmonary arteries and pulmonary veins, which bypass the normal capillary bed and cause a low resistance right-to-left shunt with refractory hypoxemia.[1] Generally, PAVMs were congenital, most commonly associated with HHT in 80% to 95%.[2,3] HHT or also known as Osler-Weber-Rendu syndrome is an autosomal dominant disease with almost complete penetrance an variable phenotype, linked to the suppression of the inhibition of *tranforming growth factor beta* (TGF beta) on angiogenesis, by mutation of the *ENG* or *ACVRL 1* genes.[4,5] A number of others conditions, are more rarely associated with acquired PAVMs, such as hepatic cirrhosis, shistosomiasis, mitral stenosis, and metastatic thyroid carcinoma etc.[3,6] PAVMs can be either simple, or complex, and isolated or multiples. The simple type (80% of cases) has a single feeding segmental artery and single draining vein and the complex type (20% of cases) has 2 or more feeding arteries or draining veins.[7,8] In our study, it was a simple AVM with right pulmonary artery communicating directly with ipsilatéral inferior pulmonary vein. The incidence of PAVM is 2 to 3 per 100 000 population and sex ratio H/F varies from 1: 1.5 to 1.8.[1,7] The age of diagnosis is very variable, range neonatal to adulthood mostly diagnosed in the first 3 decades of life.[3,7,9] Generally, clinical manifestations occur later in life.[2,4,7] Symptoms of the PAVM depend on mostly size and less on number of lesions. Solitary PAVM smaller than 2 cm, are most commonly asymptomatic.[2] Symptoms related to PAVMs found on initial assessment included dyspnea, cyanosis, digital clubbing, hemoptysis etc.[1,2,4,7] Diagnostic of PAVMs with HHT based on Curacao criteria in which, 3 of the following 4 criteria are needed for diagnosis: (1) spontaneous and recurrent, (2) epistaxis, (3) telangiectasia; (4) family history and pulmonary, cerebral, liver, spinal, and gastrointestinal arteriovenous malformation.[10] None of the criteria described above were found in our case. All patients with possible or confirmed HHT should be screened for PAVMs. PAVMs occur in about one third of patients with HHT. Children with possible or confirmed HHT should be screened for lung, brain, liver AVMs.[1,9] PAVMs may increase in size and cause a variety of life-threatening complications, such as cardiac failure, stroke cerebral, pulmonary hemorrhage, hemothorax, hemoptysis, and rupture.

On imaging, chest X-rays showed a round or oval sharply defined mass uniformly increased density. However, chest X-rays are insufficient for the diagnosis because PAVMs lesions can be misdiagnosed as tumor of pneumonia or others.[1,2,8] Computed tomography (CT) pulmonary angiography remains a reference exam to confirm the diagnosis with a sensitivity $>97\%$ and show as serpiginous of vascular nature or sharply defined nodular mass most often localizated in the lower lung lobes. Three-dimensional (3D) helical CT is also being used for the diagnosis of PAVMs.[1,2,8] However, angiography was better able to determine the angioarchitecture of individual of PAVMs than CT but it is an invasive technique and with risks, making the examination less accessible.[2,7] Echocardiography usually shows normal intracardiac anatomy.[9] Magnetic resonance imaging is less efficient than the scanner in PAVMs, and therefore less used. But it can be very useful in cerebral and hepatic AVMs in the context of HHT.[7]

The traitement of PAVM is indicated in patients with significant symptoms or complications require invasive

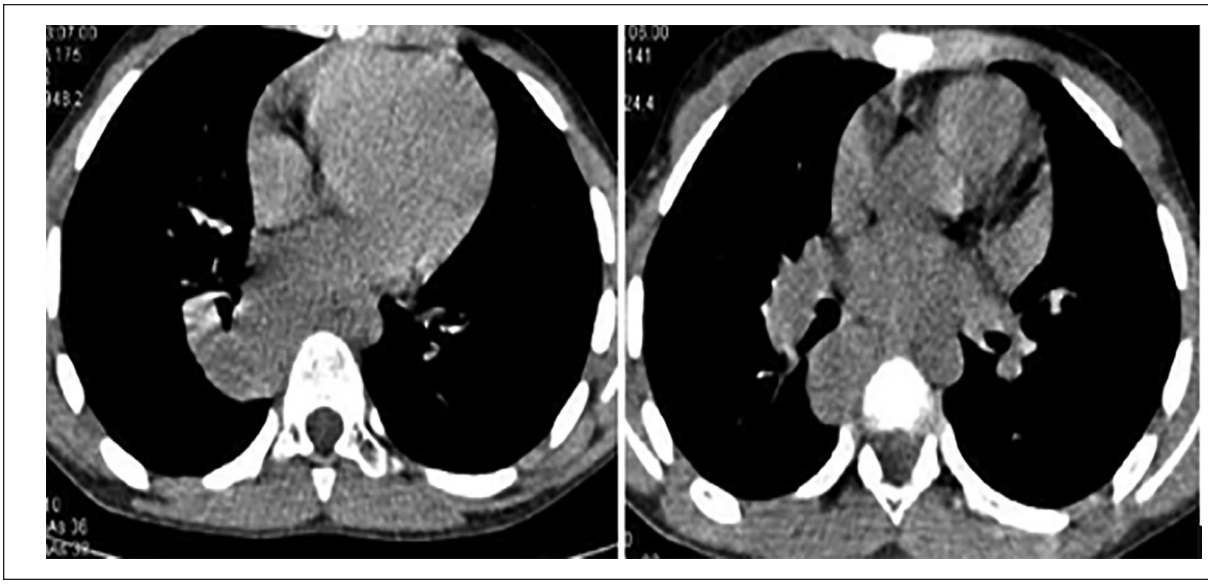

Figure 2. Axial CT without injection showing a large soft tissue oval, proximal as vascular nature in the postero-basal of the right upper lob.

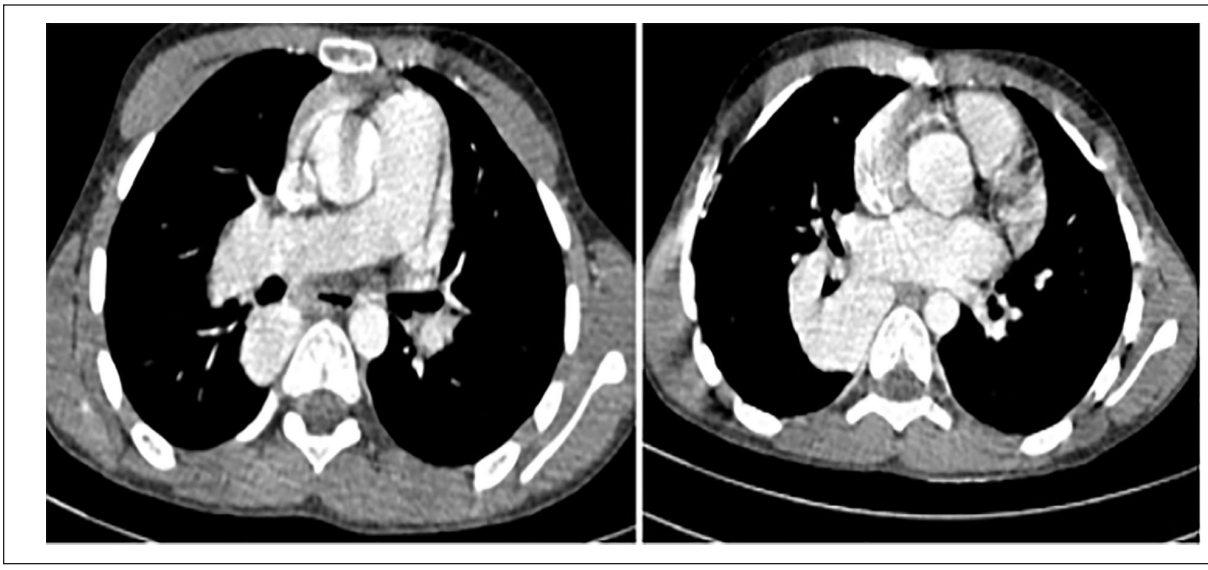

Figure 3. Axial CT confirmed with injection showing a simple large serpiginous right pulmonary arteriovenous fistula. The right pulmonary artery communicating with the right inferior pulmonary vein.

traitement. Initially, surgical option was the only option and consisted of a lobectomy until the introduction of endovascular embolotherapy.[5,8] Percutaneous endovascular treatment is less invasive and allow a shorter hospital stay but is not without risks. Embolization may be complicated by prosthesis migration during the procedure, allergy to the contrast product, and complication at the puncture site. Surgical treatment such as a lobectomy was indicated when embolization was unsuccessful or technically not feasible.[1,5,8]

It is recommended in patients with PAVMs to screen every 3 to 5 years, if a pulse oximetry test result is 97% or higher. If a pulse oximetry result test lower than 97%, or child is short breath, additional tests or treatment may be required.[2,10]

Conclusion

PAVMs are malformations most often of congenital origin, the clinical manifestations of which often appear

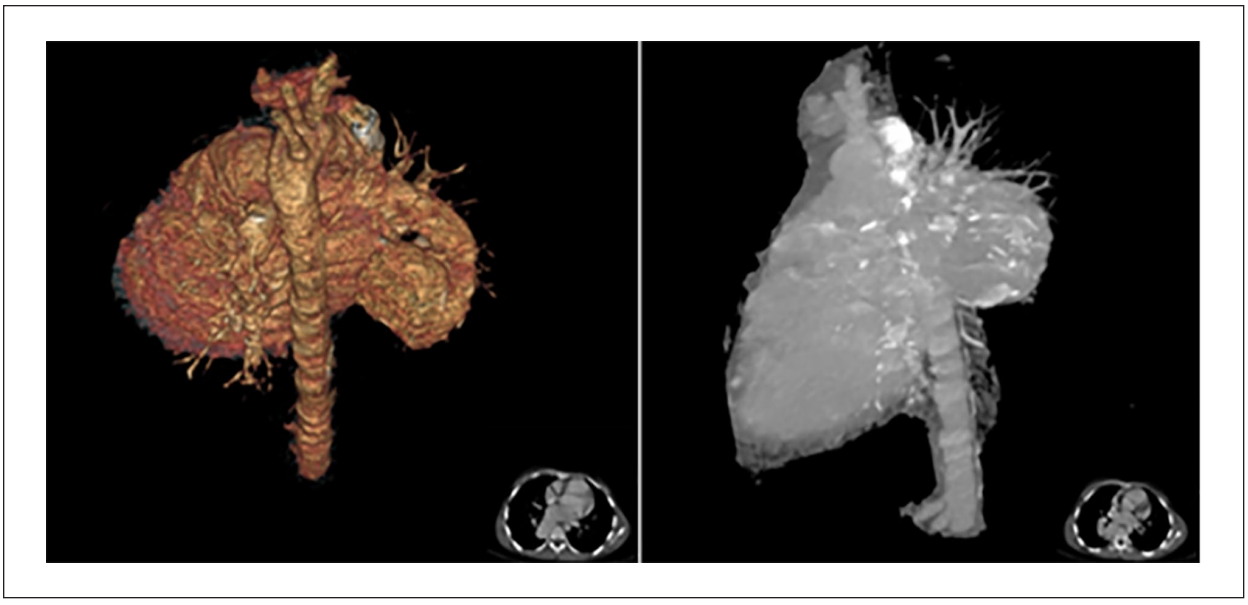

Figure 4. Dynamic 3D bone reconstruction in color and white.

late. They can be isolated but most associated with Osler-Weber-Rendu syndrome. We should think about it in front of low oximetry, short breath in the child, unimproved pneumonia, and especially in the presence of HHT. All patients with HHT and patients with suspected should be to screen by CTA and echocardiography to decide on the therapeutic choice.

Author Contributions

All authors contributed equally in this work.

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