Case Report

Pulmonary Arteriovenous Malformation: A Rare Cause of Right to Left Shunt.

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Abstract

Pulmonary arteriovenous malformations are rare anomalies where pulmonary arteries and veins communicate by passing the pulmonary capillary bed. They are mostly congenital in origin, however sometimes may be acquired. They may present as a diagnostic dilemma in plain radiography. Computed tomography angiogram has emerged as the diagnostic modality of choice not only for diagnosing the case but also for delineating the angioarchitecture that is useful in treatment planning by surgery or transcatheter embolisation. Here we present a case of a child with large pulmonary arteriovenous malformation being treated as pneumonia and finally diagnosed on computed tomography angiogram.

Keywords: Computed tomography, Pulmonary arteriovenous malformation, Right to left shunt

Introduction

Congenital heart disease with right to left shunt is a common cause for persistent hypoxemia in a child. Patient may present with right to left shunt and a normal echocardiography in case of pulmonary arteriovenous malformation (AVM) which causes shunting of blood from pulmonary artery to pulmonary vein bypassing the oxygenation in the lung capillaries.[1] The clinical presentation in such case will be similar to congenital heart disease with right to left shunt. Diagnosis is usually made during contrast enhanced echocardiography done with strong suspicion of pulmonary AVM or computed tomography (CT) of chest.[1, 2] Here we present a case with persistent hypoxemia with diagnostic dilemma, diagnosed to be pulmonary AVM on contrast enhanced chest CT.

Case Report

We present a case of a 33-month male child being investigated for fever for 3 days, dry cough for 2 days and fast breathing for 1 day. The patient's examination findings were tachypnea, peripheral cyanosis and decreased breath sound on right lower zone with crepitations. The patient's oxygen saturation was maintained at 80% with supplemental oxygen. A provisional clinical diagnosis of pneumonia with cyanotic heart disease was made; chest radiograph and echocardiography were ordered. Chest radiograph of the patient showed a round radiodense shadow in the right lower zone having sharp margin with the lung parenchyma. (Figure 1)

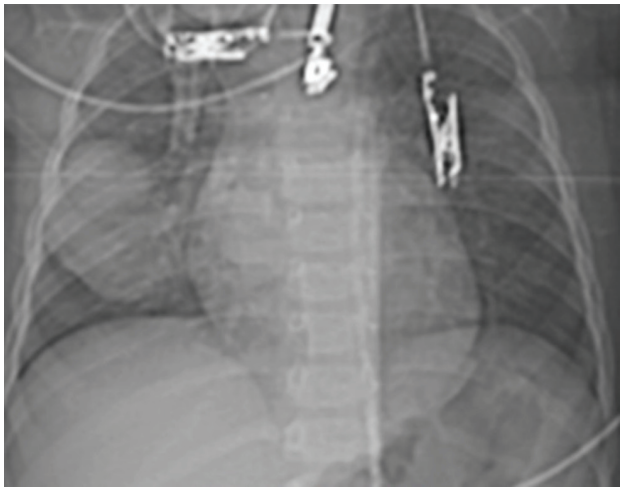

Figure 1 Radiograph showing a large well-defined lobulated lesion in right middle and lower zone. Multiple wires and tubes are also noted in the lung field.

Echocardiography was normal. The clinician made a provisional diagnosis of pneumonia and continued treatment with antibiotics. Despite treatment, the patient's saturation failed to improve and the radiographic abnormality persisted without interval change. Contrast

enhanced CT scan was ordered to further characterize the opacity. CECT chest demonstrated 4x3x3cm lobulated soft tissue density mass in the lower lobe superior segment of right lung. No calcification was noted within the lesion. The lesion showed strong heterogeneous enhancement comparable to blood pool after IV contrast administration. The right descending pulmonary artery was dilated and was seen to supply the lesion. The lesion drained into the right inferior pulmonary vein, which was also dilated. (Figure 2, 3) Rest of the lung fields appeared normal. Heart and mediastinum appeared normal. A diagnosis of large pulmonary arteriovenous malformation was made.

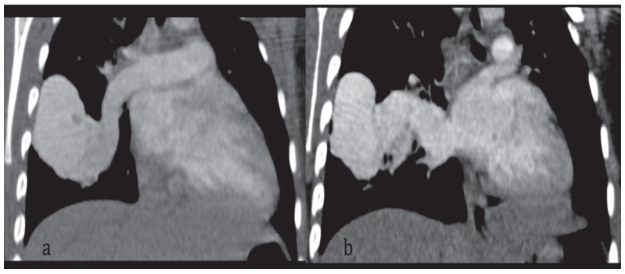

Figure 2 Contrast CT Coronal Oblique MPR images demonstrating dilated right descending pulmonary artery supplying the lesion (a) and dilated right inferior pulmonary vein draining the lesion(b).

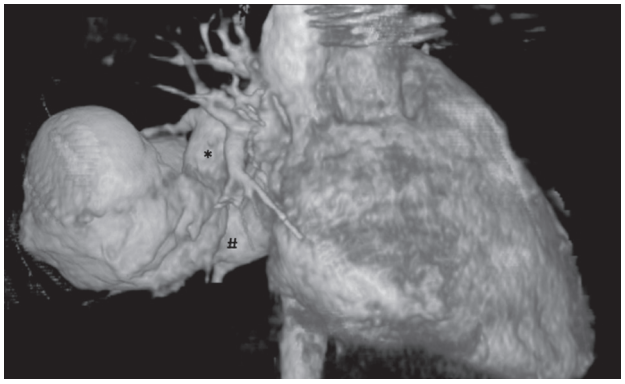

Figure 3. Volume rendered image demonstrating large pulmonary AVM supplied by descending branch of right pulmonary artery(*) and draining into right inferior pulmonary vein(#).

Discussion

An AVM is an abnormal tangle of vessels where arteries shunt directly into veins bypassing the capillaries. An AVM of pulmonary circulation is a rare occurrence. Most of the cases are congenital and in about 70%, it is associated with hereditary hemorrhagic telangiectasia. [1-3] The feeding artery is pulmonary artery in 95% of AVMs, however occasionally, it may receive feeders from systemic circulation. The drainage is to the left atrium; however anomalous drainage to inferior venacava or innominate vein has been reported[2].

The clinical manifestations of pulmonary AVMs are due to shunting of blood from right side (pulmonary artery) to left side (left atrium) of heart. The right to left shunt can lead to cyanosis if large shunt is present. Also since particles are not filtered through the shunt, paradoxical embolus can occur to systemic circulation. Pulmonary AVM usually tend to occur in the lower lobes (50-70%)[1], thus may lead to orthodeoxia (increased flow to lower lobe on upright position and thus increased shunting) and if the shunting is significant, it can cause platypnoea.

Only about 10% of the pulmonary AVM manifest during infancy and childhood. The manifestations during early age range from asymptomatic to dyspnoea and cyanosis. Bleeding manifestations (epistaxis, mucosal purpura) may be the presenting symptoms in patients with hereditary hemorrhagic telangiectasia. Complications may be due to paradoxical emboli (transient ischemic attack, stroke, migraine, brain abscess), due to hypoxemia, due to rupture of AVM leading to massive hemoptysis and hemothorax.[2]

Chest radiograph is the initial evaluation tool for evaluating pulmonary AVM. They present abnormal chest radiographs in 98% of cases as round to oval sharply defined lesions 1-5cm in diameter with sharply defined margin to lung parenchyma is present.[1, 2] Vessels from hilum may be seen connecting to the pulmonary AVM. Calcification is absent in pulmonary AVM. However these findings are nonspecific for AVM.[4]

Multidimensional computed tomography (MDCT) and computed tomography angiography (CTA) are useful in confirming the diagnosis as well as in defining the angioarchitecture and multiplicity of these lesions; also are utilized in the follow-up of treated as well as smaller untreated lesions. Ultrafast contrast enhanced CT has shown to be more sensitive than conventional angiogram to detect pulmonary AVM and is better in defining the architecture of the lesion and the feeders and draining vessels.[1, 5-7]

AVM with feeding artery more than 3mm are at increased risk of paradoxical embolism and a recommendation for treatment of pulmonary AVM either by surgery or transcatheter embolisation (TCE).[1, 5, 6] Pulmonary AVMs may grow in size if left untreated and increase the right to left shunting resulting in hypoxemia. They have also been reported to regress spontaneously after pregnancy.[6] Asymptomatic pulmonary AVMs with feeder can just be followed up by CT scan[5].

The presentation in the case presented is classical to pulmonary AVM with dyspnoea and hypoxemia at presentation. However clinical presentation is non-specific for pulmonary AVM and due to its rare occurrence, pulmonary AVM was not suspected clinically. The radiograph showed a round radiodense shadow which was not typical of pneumonia. The CECT demonstrated a large AVM with the right descending pulmonary artery as the feeder and the right inferior pulmonary vein as the draining vessel, both of which were dilated and tortuous. Although pulmonary AVMs are rare, they need to be included in differential diagnosis when nodule is seen in chest radiograph with a suggestive clinical history. Missing to evaluate such a nodule in chest radiograph can cause catastrophic event to the patient.

Conflict of interests: None declared.

Reference

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