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"CT Plethora" sign in high resolution computed tomography (HRCT) of diffuse pulmonary arteriovenous malformation (PAVM)

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Abstract

Diffuse PAVM is rare and poses significant morbidity and mortality. It is an important differential diagnosis in patients with hypoxia and clubbing of unknown cause. Diffuse PAVM lacks visible nidus, as these are microscopic channels that shunt the arteries to the veins with absence of any capillary bed. Thus the CT images will not show the classical nodular nidus of an arteriovenous malformation as the lesions are diffuse and microscopic. Instead, a reconstructive HRCT shows diffuse increase nodular vasculature of bilateral lungs; resembling pulmonary plethora classically described in chest radiography. We report a case with diffuse PAVM in a child who presents with central cyanosis.We suggest that reconstructive HRCT finding of "CT plethora"as a useful sign for non-invasive diagnosis of diffuse PAVM.

Keywords

diffuse pulmonary arteriovenous malformation; high resolution computed tomography; hrct; computed tomography angiogram; diffuse pavm; congenital

Abbreviations

ECHO: Echocardiography; HRCT: High resolution computed tomography; CTA: Computed tomography angiogram; PAVM: Pulmonary arteriovenous malformation; MIP: Maximum intensity projection

Introduction

Diffuse Pulmonary Arteriovenous Malformation (PAVM) is rare but is an important subset of the PAVM population. It poses significant morbidity and mortality [1].

Case Report

A 4 year old child presented with recurrent bronchopneumonia and noted to have central cyanosis with oxygen saturation at room temperature between 77- 80% and clubbing but no telangectasia. Clinically, he is a small boy with no dysmorphism. BP 88/60, HR 123bpm. Auscultations of the lungs was clear. Hyperoxia test and chest radiography were done and found to be normal. Methemoglobin and haematocrit were 1.3% and 47. Arterial blood gases shows pH 7.47, pCO_2 28.2, p02 67mmHg, BE -2.7, HCO_3 23 and lactate 2.5. Echocardiogram (ECHO) shows normal heart structures with no features of pulmonary hypertension. However, contrast bubble ECHO test was positive and suggestive of pulmonary AVM. High Resolution reconstruction Computed Tomography (HRCT) with maximum intensity projection at slab thickness of 30, from a Computed Tomography Angiogram (CTA) performedshowed prominent diffuse bilateral peripheral network of vessels.

Discussion

In PAVM, there is direct communications between the pulmonary arteries to the veins, bypassing the capillary system. The aetiology is congenital; due to failure of differentiation of the embryonic vascular plexus. PAVM may be solitary and discrete or diffuse [2]. Patients usually present with hypoxia [3]. Embolization therapy does not improve the profound hypoxia [3] in patient with diffuse PAVM [3]. They are also at increased risk of neurologic complications due to paradoxical emboli causing strokes and cerebral abscesses.

Diffuse PAVM is associated with hereditary haemorrhagic telangiectasia (HHT) [4,5] and are more commonly bilateral. Diffuse bilateral PAVM is very rare, reported less than 20 times since 1930s [2-7]. There is a female predilection in bilateral diffuse PAVM [1].

To establish a CT diagnosis of a classical PAVM, it is necessary to show feeding and draining vessels extending to an AVM nidus on a Computed Tomography Angiogram (CTA). However, in the diffuse subset this is not the case as the channels shunting the arteries to the veins are microscopic and diffuse. Hence, the only CT finding will be increase in the vasculature till the lung periphery. Our observations, convinced us that the appreciation of this findings are real and the appearance are not seen in normal HRCT. Since CTA is part of the workout for suspected pulmonary AVM, the reconstruction of the CTA data into high resolution CT could easily be done.

Diagnosis of diffuse PAVM is made from contrast ECHO, angiogram and biopsy. Use of CT is not well established in diagnosis of diffuse PAVM. Furthermore CTA thorax or HRCT for these are usually are normal from all the previous case report [8]. From our observation in this case and from the reviews of previously published case reports; there is increase nodular vasculature or pulmonary plethora on HRCT best seen on the thick slab (we suggest 30 slab thickness) maximum intensity projection (MIP). Pulmonary plethora is a radiographic finding in chest radiographs to described increase numbers of lung vessels. A Korean case report similarly discussed of role of maximum intensity projection in the diagnosis of diffuse PAVM [7]. The paper suggests that CTA with MIP reconstruction will reveal subpelural nodules. Our case shows increase

nodular vasculature throughout the lung parenchyma. Thus, we recommend HRCT with MIP reconstruction to be performed and "CT plethora" sign (see image 1a) to be used to prompt radiologist and clinician of the diagnosis of diffuse PAVM. Image 1b shows the normal lung vasculature as a comparison.

HRCT is non-invasive as appose to conventional angiogram or lung biopsy. The contrast ECHO on the other hand is operator dependent and the contrast agent is expensive. Furthermore, HRCT may give additional information on lung parenchyma, gross heart structure and pleural effusion.

Conclusion

"CT plethora" sign in reconstructive HRCT is useful as a non-invasive diagnosis of diffuse PAVM.

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Figures

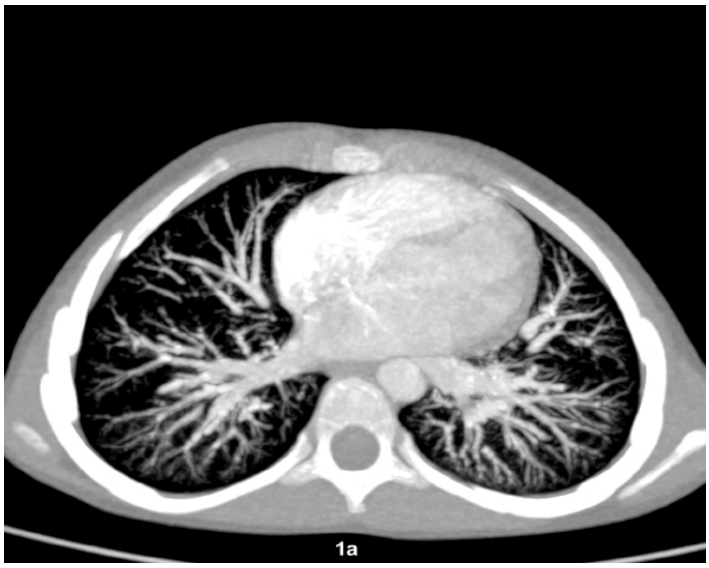

Figure 1a: Axial CTA image with thick slab (30) Maximum intensity projection(MIP) of the lungs in diffuse PAVM shows increase nodular vasculature suggestive the "CT plethora" sign.

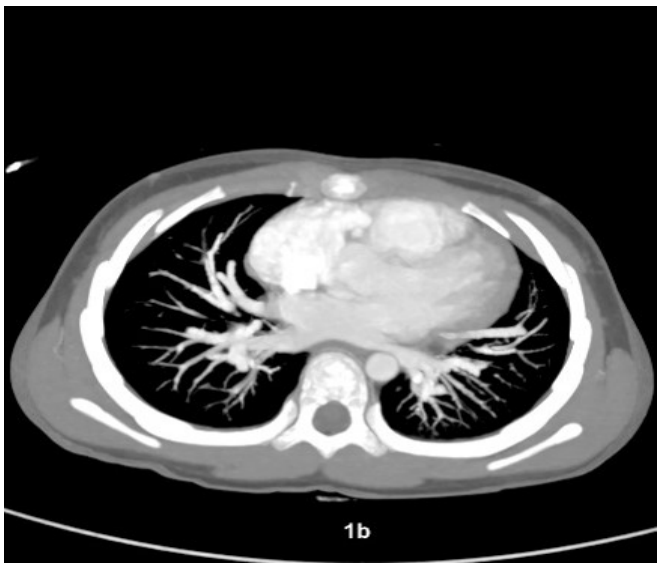

Figure 1b: Axial CTA image with thick slab (30) Maximum intensity projection (MIP) of normal intraparenchymal pulmonary vessels.

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