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Case Report

If You See a Round Density on Chest X-ray in a Cyanotic Child, Suspect Pulmonary Arteriovenous Malfomation

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Abstract We reported a 15-month-old infant who presented with dyspnoea and cyanosis. She was hospitalised with the diagnosis of bronchopneumonia but the patient showed no clinical improvement with antibiotic treatment. There was persistent cyanosis and there was progressive radiological change on anterior-posterior chest X-ray (CXR) during follow-up. A round density was noted on the CXR, tests were performed to exclude pulmonary arteriovenous malformations (PAVM) and the diagnosis was made. Transcatheter embolisation of the pulmonary arteriovenous malformation was performed with an Amplatzer Vascular Plug. In patients with suspected pneumonia resistant to treatment, presence of round or oval density on CXR should raise the suspicion of PAVM.

Key words Cyanosis; Pulmonary arteriovenous malformation; Round density

Introduction

Pulmonary arteriovenous malformations (PAVM), are abnormal direct connections between the pulmonary artery and pulmonary veins, which can cause right to left shunt.[1] They are also known as pulmonary arteriovenous fistula, aneurysm, pulmonary angioma, cavernous haemangioma or telengiectasis.[2] The incidence in the general population is estimated to be 2-3/100,000. The majority of cases are seen in the 5th-6th decades of life, and only 10% are seen in children.[1] We reported a paediatric patient who presented with dyspnoea and cyanosis, unresponsive to the treatment for pneumonia, and during follow-up PAVM was diagnosed.

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Case Report

A 15-month-old female infant presented at another centre with shortness of breath and cyanosis, was referred to our hospital with the presumptive diagnosis of congenital heart disease and bronchopneumonia. On admission, the patient had intermittent cough and was afebrile. Respiratory rate was 35/min and saturation was 80%. Air entry were reduced in the basal region of right lung with no rales nor rhonchi. There was no finger clubbing nor telengiectasis. Congenital heart disease was excluded by telecardiography, electrocardiography, echocardiography and arterial blood gas examination results.

Haemoglobin was 13.4 g/dL, haematocrit as 40.2, white

blood cells as 14,900/mm^3 (predominantly fragmented), and C-reactive protein as 3 mg/l. On the anterior-posterior chest X-ray (CXR), there was infiltration in the inferior lobe of the right lung (Figure 1a). As the patient showed no clinical improvement with the initial treatment of sulbactam ampicillin, with a presumptive diagnosis bronchopneumonia, this was changed to vancomycin, meropenem, clarythromycin. The cyanosis continued and progression was observed on CXR during follow-up (Figure 1b). On thoracic computed tomography (CT), there was an intense contrast involvement consistent with vascular malformation, and a dilated pulmonary vein draining to the left atrium (Figure 1c). Pulmonary angiography was done and a diagnosis of right PAVM was made (Figure 1d).

Patient was referred to a tertiary referal centre where transcatheter embolisation of PAVM with an Amplatzer Vascular Plug was done successfully. After the procedure, cyanosis resolved, saturation was 97% in room air and the radiological opacity on the CXR resolved (Figure 1e). On follow-up examination when the patient was 18 months old, parents complained that patient had squint and reduced vision. Magnetic resonance imaging examination the brain was normal. However orbita tomography examination showed a soft tissue density mass with calcification measuring 1.1x1.1x0.7 cm in the posterolateral of the left globe. A diagnosis of retinablastoma was made. After 6 cycles of chemotherapy, there was significant shrinking in the lesion in the posterolateral of the left globe on the follow-up orbital CT.

Discussion

PAVM is rarely seen in children. The aetiology and prognosis are not well known. It can be due to congenital cause, acquired after cardiac surgery (Glenn or Fanton procedures) hepatic cirrhosis, infections, amyloidosis, mitral stenosis, chest trauma, and bronchiectasis. PAVMs may be simple or complex, single or multiple. Eighty percent of the cases are simple PAVM with a single feeding

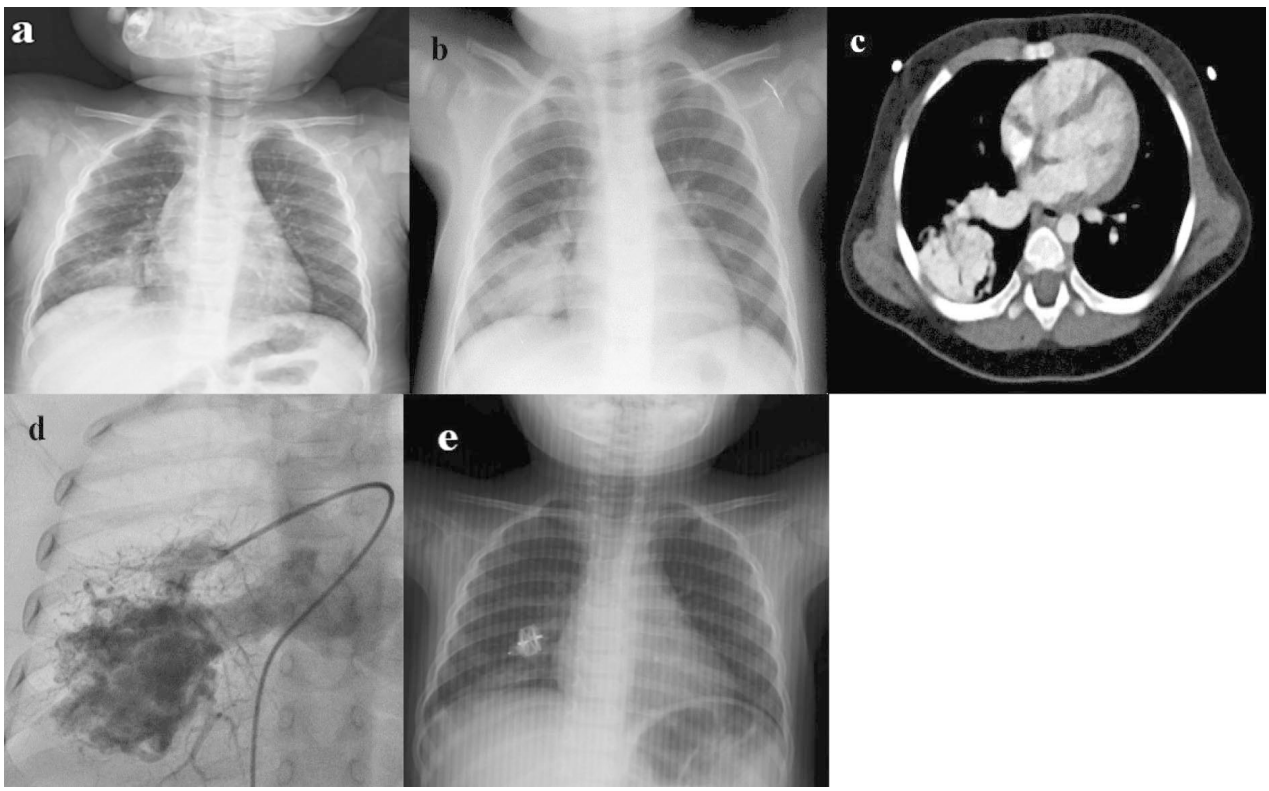

Figure 1 (a) Anterior-posterior chest X-ray at the time of presentation; infiltration in the inferior lobe of the right lung. (b) Radiograph on the 7th day of treatment; increased density in a round form 3-4 cm in diameter in the inferior basal of the right lung. (c) Thoracic CT; intense contrast involvement consistent with vascular malformation and dilated pulmonary vein draining to the left atrium. (d) Pulmonary angiography; intense contrast involvement consistent with PAVM and draining vein below. (e) Follow-up anterior-posterior chest X-ray following the application of Amplatzer Vascular Plug.

segmental artery and single draining vein. In complex type there are two or more feeding arteries or draining veins. PAVM with autosomal dominant Hereditary Haemorrhagic Telengiectasis is present in 36% of single lesions and 57% of multiple lesions.[3] The diagnosis of HHT could not be made in this case as there was no family history, epistaxis, telengiectasis or visceral organ involvement. No other causes of PAVM could be found. Therefore this is a case of simple congenital PAVM.

More than half of cases with PAVM are asymptomatic, and lesions are often dedected by routine CXR. Common presenting symptoms are listlessness, dyspnoea without exercise, chest pain, haemoptysis, and recurrent epistaxic attacks. In the majority of patients (75%) there are no abnormal physical examination findings. Cyanosis, finger clubbing, telengiectasis and systolic murmur may be found in the physical examination. The classic triad of PAVM is cyanosis, finger clubbing and polycythemia, but this is only present in 20% of cases. Our patient presented with cyanosis without any finger clubbing, telengiectasis and heart murmur.

With increasing age, necrosis may be seen in the vessel wall as a result of progressive growth in the PAVM. This may cause an increase in the amount of shunting or haemorrhage. In addition, these lesions are associated with several life-threatening complications which include heart failure, stroke, cerebral abscess, pulmonary haemorrhage, haemothorax, and rupture. These complications can develop at any age regardless of the lesion size.[1] The patient did not have any the above complications. Interestingly, retinablastoma was diagnosed during follow-up of the patient.

The diagnosis of PAVM is made through a combination of clinical presentation as well as diagnostic imaging tests such as CXR, thorax CT, transthoracic contrast echocardiography, and pulmonary angiography. CXR abnormality is present in approximately 98% of PAVM cases.[1] The classic radiological appearance is a round or oval shaped opacity often in a lobular patern with sharp borders usually in the inferior lobes and around 1-5 cm in diameter.[4] This radiological abnormality can lead to a diagnosis pneumonia or tumour. Alternatively this can present as parenchymal haemorrhage or atelectasia associated with bronchi compression. Our patient presented with cough with no spesific radiological abnormality. Patient was treated as pneumonia. However patient was unresponsive to treatment and repeated CXR showed a round opacity, PAVM was suspected and diagnosis was made.

Treatment of PAVM can be either endovascular embolisation therapy or surgical excision. In endovascular embolisation therapy, embolisation coils or mini-balloons can be used. Other potential therapy include cryotherapy, radiotherapy or laser therapy. Although transcatheter embolisation therapy is frequently used to treat PAVM, potential problems and complications include persistence of the lesion, growth of new feeding arteries, migration of the embolic agent, haemorrhage, tachyarrhythmia, bradyarrhythmia, air embolisation and pulmonary hypertension can occur.[5] Our patient with suspected PAVM, CXR, thorax CT and CT-pulmonary angiography were utilised to further evaluate the lesion. Echocardiography is performed to exclude pulmonary hypertension. If there is pulmonary hypertension, significant shunts, giant lesions involving the whole lobe, renal failure or other specific conditions. (e.g. intravenous contrast dye allergy, failed endovascular application), surgical excision is indicated.[4] Our patient was treated with transcatheter Amplatzer Vascular Plug and no complications were observed during the follow-up period.

Conclusion

PAVM can cause significant morbidity and mortality. Early diagnosis and treatment are important to prevent complications. In patient with cyanosis resistant to treatment PAVM should be kept in mind if there is round or oval opacity seen in CXR.

Declaration of Interest

There is no conflict of interest to declare.

References

1. Khurshid I, Downie GH. Pulmonary arteriovenous malformation. Postgrad Med J 2002;78:191-7.
2. Biçakçioğlu P, Gülhan SŞ, Sayilir E, et al. Surgical treatment of pulmonary arteriovenous malformations. Turk J Med Sci 2017; 47:161-6.
3. Remy J, Remy-Jardin M, Wattinne L, Deffontaines C. Pulmonary arteriovenous malformations: Evaluation with CT of the chest before and after treatment. Radiology 1992;182:809-16.
4. Dokumcu Z, Ozcan C, Alper H, Erdener A. Pulmonary arteriovenous malformation in children. Pediatr Int 2015;57:708-11.
5. Faughnan ME, Thabet A, Mei-Zahav M, et al. Pulmonary arteriovenous malformations in children: Outcomes of transcatheter embolotheraphy. J Pediatr 2004;145:826-31.