An unusual congenital pulmonary arterio-venous fistula

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Abstract

Pulmonary arteriovenous malformations (AVM) are rare causes of a cardiac murmur in the paediatric population. They are caused by abnormal communications between pulmonary arteries and veins that are most commonly congenital in nature. Although these lesions are fairly uncommon, they are an important differential diagnosis to consider in patients with common pulmonary problems such as hypoxaemia and/or a pulmonary nodule(s). This report illustrates the clinical presentation, radiological features and pathological findings in an eight-month-old boy.

Key words
Fistula, arteriovenous, congenital, trisomy 21

Case report

An eight-month-old male patient with Down's syndrome was referred for cardiac catheterisation at the Red Cross Children's Hospital. He had signs of a left-to-right shunt with bounding pulses and a machinery murmur at the upper left sternal border. Both on clinical grounds and on echocardiographic examination it was felt that he had a PDA. Cardiac catheterisation (with a view to possible coil embolisation) was performed. At catheterisation, a duct was seen but was noted to be small. The larger feeding vessel was noted during the same angiogram. A contrast CT scan confirmed that there was a pulmonary arteriovenous malformation occupying the lower lobe of the left lung (Figures 1, 2 and 3). An artery originating from

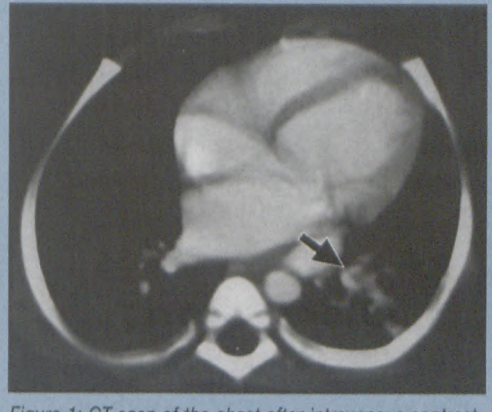

Figure 1: CT scan of the chest after intravenous contrast administration demonstrates a brightly enhancing conglomerate of vessels (arrow)

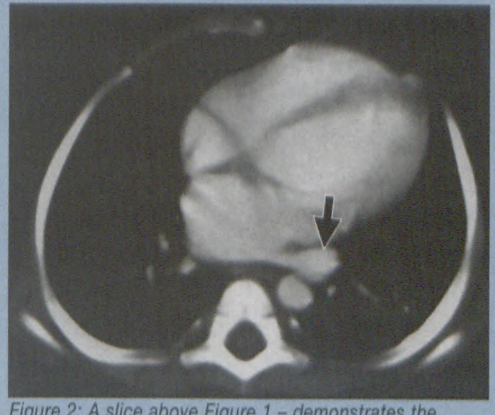

Figure 2: A slice above Figure 1 – demonstrates the vascular malformation draining into the left atrium via a large draining vein (arrow)

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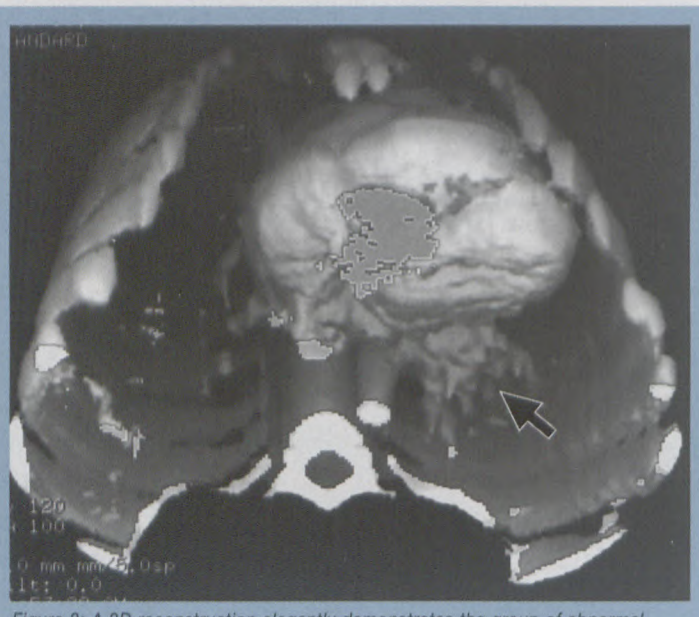

Figure 3: A 3D reconstruction elegantly demonstrates the group of abnormal vessels (arrow) and its arterial blood supply from the descending thoracic aorta

the descending aorta, just above the diaphragm, supplied this lesion, which was seen to drain into the left atrium. The feeder vessel was ligated and a left lower lobe lobectomy was performed. This showed a vascular malformation composed of large, dilated interconnected vessels, many of them histologically resembling arterialised veins (Figure 4).

Discussion

Since their first description at autopsy in 1897,[1] these lesions have also been called pulmonary arteriovenous "fistulae", pulmonary arteriovenous "aneurysms", "hemangiomas" of the lung, "cavernous angiomas" of the lung, pulmonary "telangiectases" and pulmonary arteriovenous malformations.[2] The term "pulmonary arteriovenous malformations" (PAVM) appears to be most widely accepted in modern literature.[3]

Although most commonly a congenital abnormality, abnormal communications between blood vessels of the lung may also be found in a variety of acquired conditions, such as hepatic cirrhosis[4] and bronchiectasis[5] and should always be considered in the management of such patients.

PAVMs occur twice as often in women as in men, but there is a male predominance in new-borns (as in the index case), where symptoms may vary from being totally absent to severe with cyanosis, congestive heart failure and even fulminant respiratory failure.[6]

Around 10% of cases of PAVM are identified in infancy or childhood, with a peak incidence occurring in the fourth to sixth decades of life. Approximately 70% of the cases of PAVM are associated with hereditary haemorrhagic telangiectasia (HHT), an inherited disorder of autosomal dominant inheritance, characterised by arteriovenous malformations of the skin, mucous membranes and visceral organs.[7] Patients with a PAVM should therefore be screened for this syndrome. There were no such features in our patient, despite an extensive search that included contrast CT scan of the brain.

The classic clinical triad of dyspnoea, clubbing and cyanosis is rarely seen, with adult patients presenting most commonly with epistaxis, reflecting the strong association with HHT. Dyspnoea and haemoptysis are also common symptoms and in half of cases a bruit or murmur can be heard, most audible during inspiration.[3] Our patient had a particularly loud murmur that was clinically thought to be a large PDA.

PAVM usually occurs in the lower lobes and is solitary in 75% of cases.[8] It can be classified as either simple or complex. The *simple* type (80-90% of cases) is defined as having a single feeding segmental artery and a single draining vein. The rest are complex, with two or more feeding arteries or draining veins.[9]

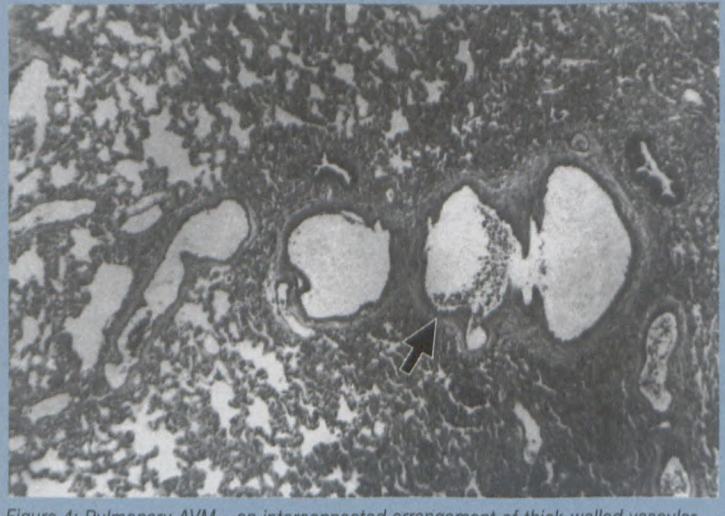

Figure 4: Pulmonary AVM – an interconnected arrangement of thick-walled vascular channels (arrow) within the lung parenchyma (H&E x 4)

In the majority of patients (about 95%), the AVMs are supplied by pulmonary arteries. AVMs are supplied by systemic arteries less frequently.[10]

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Such AVMs need to be differentiated from true sequestrations. Drainage is usually to the left atrium, but anomalous drainage to the inferior vena cava or innominate veins has been reported.[2,11]

Pathological examination shows that PAVMs are similar to AVMs occurring elsewhere in the body. The malformations may have one of three typical appearances: (1) a large, single sac, (2) a plexiform mass of dilated vascular channels, or (3) a dilated and often tortuous direct communication between artery and vein.[9,11] Mural thrombi or calcifications are also occasionally seen.[2]

The classic roentgenographic appearance of a PAVM is that of a round or oval mass of uniform density, frequently lobulated but sharply defined and more commonly in the lower lobes.[3] Although uncommon, multiple lesions may be identified.[12] Solitary PAVMs will often show feeding vessels on chest radiography, with the artery radiating from the hilus and the vein deviating towards the left atrium.[13]

Despite advances in diagnostic techniques mentioned thus far, contrast pulmonary angiography remains the gold standard in the diagnosis of PAVM. Contrast echocardiography, computed tomography, radionuclide perfusion lung scanning, pulmonary angiography and magnetic resonance imaging are all further useful modalities.[3]

Treatment depends on the clinical symptoms, signs and size of the lesion and includes surgical resection, embolisation therapy or hormonal manipulation.

In summary, therefore, PAVMs are uncommon paediatric problems, but should be considered in patients with (1) one or more pulmonary nodules, (2) mucocutaneous telangiectases and (3) unexplained clinical findings such as dyspnoea, haemoptysis, hypoxaemia, a machinery heart murmur, clubbing or cyanosis.

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A review of paediatric abdominal masses

Conclusion

Any abdominal organ may develop a mass. The spectrum of clinical presentations is broad and assessment of the child with an abdominal mass may, at first, seem daunting. A small proportion of entities account for the vast majority of cases and a knowledge of the statistical distribution, age and sex of the patient, clinical presentation (notably pain or pyrexia) and imaging characteristics (especially solid vs cystic nature) allows the formulation of a focused differential diagnosis.

Ultrasound has proved to be an invaluable tool in the assessment of paediatric abdominal masses because it is safe and efficacious, but the diagnostic capabilities of CT may often outweigh the radiation risk, especially in the older child. MRI is increasingly more useful and may be the modality of choice in the future.

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