

DIFFUSE PULMONARY ARTERIOVENOUS MALFORMATION: UNUSUAL CAUSE OF CYANOSIS IN INFANCY

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Pulmonary arteriovenous malformation (PAVM) is a rare cause of cyanosis in early infancy. The disorder usually appears in late childhood or early adult life, with dyspnea on exertion, clubbing and cyanosis. Fistulous vascular communication in the lungs may be large and localized, or small, multiple and scattered. The essential defect of a PAVM is a right-to-left shunt from the pulmonary artery to the pulmonary vein, bypassing the lungs. The clinical manifestations and the age of onset of the symptoms depend on the size of the shunt. This intrapulmonary malformation is described in a young infant who presented with severe cyanosis.

Case Report

A 45-day-old Saudi infant presented to our hospital with progressive cyanosis and tachypnea of one day. He was born at term after a normal pregnancy, and without prenatal complications. He was well until the age of 15 days, when mild cyanosis with crying and irritability was noticed by the mother.

On examination, the infant was ill-looking, with central cyanosis and tachypnea. Cardiovascular examination revealed normal-volume peripheral pulses, which were felt in both upper and lower limbs, normal first and second heart sounds and short systolic murmur grade 2/6, which were heard at the second intercostal space near the left sternal border. There was no murmur from the back or from both axillae. Chest examination revealed normal vesicular breathing, equal on both sides of the chest. The examination of abdomen, central nervous system and skin did not reveal any abnormality.

Investigations showed arterial blood gases (room air) to have pH of 7.25, PCO₂ 36 mm Hg, PO₂ 29 mm Hg, HCO₃ 14 mmol/L, and O₂ sat 41%. Chest x-ray showed normal cardiac size and diffuse reticulonodular pulmonary infiltrates in both lung fields (Figure 1).

Electrocardiogram showed sinus rhythm, normal QRS axis, and no right ventricular hypertrophy. Infection

Hyperoxic test (supplement of 100% O₂ for 10 min.) was performed, but the oxygen saturation remained at 50%, which suggested a cardiovascular cause of the cyanosis rather than a parenchymal lung disease.

The patient was admitted to the pediatric intensive care unit and connected to a ventilator for controlled ventilation. Despite the high setting of the ventilator with adequate ventilation, perfusion and supplemented oxygen (FIO₂ 98%), the oxygen saturation and the arterial PO₂ remained the same.

Arterial blood gases on ventilator showed a pH of 7.36, PCO₂ 40 mm Hg, PO₂ 34 mm Hg, HCO₃ 22 mmol/L, and O₂ sat 62%. Transthoracic echocardiography was performed, which showed normal function of both ventricles, normal heart structure, apart from a small patent ductus arteriosus (PDA) with left-to-right shunt, but did not reveal any features of pulmonary hypertension.

Contrast echocardiography study suggested the presence of intrapulmonary arteriovenous malformation with significant right-to-left shunt, as evidenced by rapid filling of the left atrium with dissolved bubbles. Right heart catheterization was performed, and the investigation was kept to a minimum because of the poor condition of the patient. Oxygen saturation measured in all pulmonary veins was significantly low (55%-58%, Table 1). The mean

TABLE 1. Hemodynamic data.

Site	Pressure (mm Hg)	O ₂ Sat %
Left lower pulmonary vein	—	54
Left upper pulmonary vein	—	56
Right lower pulmonary vein	—	58
Right upper pulmonary vein	—	54
Left atrium	9 (m=7)	43
Right atrium	7/3 (m=2)	23
Right ventricle	32/3 (m=5)	—
Main pulmonary artery	30/6 (m=15)	30
Left pulmonary artery	22/7 (m=15)	—
Right pulmonary artery	18/7 (m=12)	—
Superior vena cava	—	32
Inferior vena cava	—	25
Aorta	78/31 (m=49)	45

QP/QS: 0.92; right-to-left shunt: 29%.

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screen—blood culture, urine analysis and culture, CSF analysis and culture—was normal. Other investigations, including CBC, electrolytes, renal profile and hepatic profile, were normal.

FIGURE 1. Plain chest x-ray showing normal cardiac size and diffuse reticulonodular pulmonary infiltrates in both lung fields.

FIGURE 2. Main pulmonary artery angiography demonstrates diffuse reticular pattern on both sides of the lungs, due to diffuse bilateral AV malformations. The contrast media is seen in the left atrium immediately after injection of the dye in the main pulmonary artery.

pulmonary artery pressure (15 mm Hg) and pulmonary arteriolar resistance index (1.2 Wood's unit) were normal. The pulmonary/system flow ratio (QP/QS) was 0.92, and there was significant right-to-left shunt at lung level (30% of the right ventricular output).

Injection of contrast media in the main pulmonary artery showed immediate opacification of the left atrium and diffuse reticular pattern on both lungs (Figure 2). Selective injection of contrast media in the right and left pulmonary arteries showed similar findings.

The result of cardiac catheterization (hemodynamic data and angiography) confirmed the presence of diffuse bilateral intrapulmonary arteriovenous malformation, with significant right-to-left shunt.

From the results of investigations, including the cardiac catheterization, the options of treatment were very limited, due to extensive diffuse lesions in both lungs, and the only way to treat the patient was by lung transplant, which was not feasible. The patient expired at the age of 54 days after a cardiopulmonary arrest.

Discussion

Although single and isolated PAVMs are not uncommon, multiple, small, diffuse bilateral PAVMs are rare. The largest study was reported by Dines et al.,¹ who reviewed 101 patients with PAVM in a 30-year period. On reviewing case reports from different centers and comparing them with ours, we noticed that the more diffuse the PAVMs are, the earlier the age of presentation. Clarke et al.² reported two cases of massive PAVM presenting with cyanosis in newborn babies. Knight and Mee³ reported three cases of multiple PAVM which presented at five months, two years, and nine years of age, respectively.

The plain chest radiograph of a PAVM is usually abnormal, but often in a nonspecific way. PAVMs typically appear as peripheral, rounded noncalcified lesions with a variety of vascular communications with the hilum. A similar pattern was seen in our case, but the peripheral lesions were smaller.

The presence of a right-to-left shunt can be demonstrated using contrast echocardiography, which shows abnormal right-to-left shunt, as we found in our case.

Transcatheter coil embolization can be successful when the lesions are localized to a few lobes. Surgery (segmental resection, lobectomy) is usually successful when the lesions are large and localized to one or two lobes. Taylor et al.⁴ pioneered the technique of therapeutic embolization of pulmonary artery, while Hirota et al.⁵ have described the long-term results of percutaneous transcatheter embolization of PAVM with spring coils. Marin-Garcia et al.⁶ described an infant with PAVM who had surgical ligation of the fistulas and limited resection of parenchyma from the right lung, and was followed at 11 months of age by successful steel coil embolization of the residual fistulas. However, neither modality of the therapy were possible in our patient, due to the diffuse nature of the PAVM.

PAVMs are rare vascular structures which allow shunting of the blood between the pulmonary artery and pulmonary vein. The diagnosis of PAVM should be considered in infants with severe cyanosis without structural cardiac lesion or pulmonary hypertension, after excluding other causes of cyanosis, such as parenchymal lung disease and the rare methemoglobinemia. A high index of suspicion is required for a successful echocardiographic diagnosis of PAVM in a cyanotic infant with the use of contrast echocardiography.

Pulmonary angiography is mandatory whenever a diagnosis of PAVM is suspected, in order to confirm the diagnosis and a precise identification of the number and location of all lesions before embolization or surgical resection is undertaken. When clinical features and contrast echocardiography suggest the diagnosis, and the pulmonary angiography results are normal, open lung biopsy is required to confirm the diagnosis, in which case, the shunt may be due to multiple microscopic vessels.

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