Pulmonary arteriovenous malformation – a rare cause of hypoxemia

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ABSTRACT

We present the case of a newborn with pulmonary arteriovenous malformation, located in the left lower lobe of the lung. The venous side of the malformation was connected to a large common vein, which drained into the left atrium between the two left pulmonary veins.

The diagnosis was based on the echocardiography and angiographic computerized tomography.

She underwent left pneumonectomy at the age of 28 days. After the surgery, she developed pulmonary hypertension and needed high doses of nitric oxide and cardiac support drugs. Her general condition improved for a few days, but 14 days after surgery, she developed acidosis, severe right ventricular dysfunction, severe pulmonary hypertension, and she died at 48 days of life. We would like to emphasize the importance of a prompt detection and treatment of pulmonary arteriovenous malformation, in order to prevent serious clinical consequences.

Key words: arteriovenous malformation; hypoxemia; late diagnosis; pneumonectomy; right ventricle dysfunction

Established facts. Very rare malformation in newborns, manifested with hypoxia, cyanosis; diagnosed by angiographic computerized tomography and often associated with hereditary hemorrhagic telangiec-tasia (Osler-Weber-Rendu syndrome).

Novel insights. Normal aspect of the pulmonary x-Ray; normal echocardiography on the beginning; complicated after the surgery with severe right ventricular dysfunction.

INTRODUCTION

Pulmonary arteriovenous malformations (PAVM) are a rare cause of hypoxemia in the neonatal period. They are caused by abnormal communications between pulmonary arteries and pulmonary veins, which are most commonly congenital.

Without gas exchange it produces hypoxemia to a varying degree. Hemodynamically signi-

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ficant, the right-to-left shunt, if not recognized and treated, may be life threatening.

If the shunt is not large enough to produce clinical evidence of hypoxia, it can remain undiagnosed until the 3rd or 4th decade of life, when most of the cases become symptomatic.

CASE REPORT

A female born at 38 weeks of gestation weighing 2,500 g, with Apgar score 9 at 1 min, presented with cyanosis at three days of age. She was evaluated and admitted to a Level 3 maternity hospital, where her oxygen saturation was found to be 80-90%. Infection and cardiac malformation were ruled out. As her condition did not improve despite assisted ventilation, systemic and inhaled corticosteroids, and inhaled bronchodilator, she was referred to our hospital at 20 days of age for further evaluation and management.

The clinical picture on admission: T37.3°C, weight 2,400 g, generalized cyanosis, on conventional mechanical ventilation, oxygen saturation 75% with F_iO_2 1.0, normal pulmonary auscultation, heart rate 146 beats/min, blood pressure 95/74 mmHg, otherwise normal clinical exam.

On admission to our unit, she was on conventional mechanical ventilation, and we started empirically inhaled nitric oxide (NO). For cardiac support, she received dopamine, dobutamine, and furosemide, and for preventing infection she received antibiotherapy.

Laboratory results: normal CBC, CRP 8.8 mg/l, blood gas: metabolic acidosis.

Chest x-rays showed a normal cardiothoracic ratio, reduced pulmonary transparency in the left parahilar region and an area of increased opacity in the median third of the left lung, considered to be atelectasis.

Echocardiography: normal intracardiac relationships, and a pulmonary venous structure draining from the left lung above the heart into the left atrium. Contrast echocardiography was performed by injecting saline through a central venous catheter into the superior vena cava (SVC). This procedure demonstrated initial filling (bubbles) in the SVC and right atrium, then in 2-3 seconds into the left atrium via a venous collector that entered the left atrium between the two left pulmonary veins.

Angiographic computerized tomography (Angio CT) examination of the thorax revealed aberrant vessels – arterio-venous malformation (sizing 48 mm x 44 mm x 5 mm) in the superior

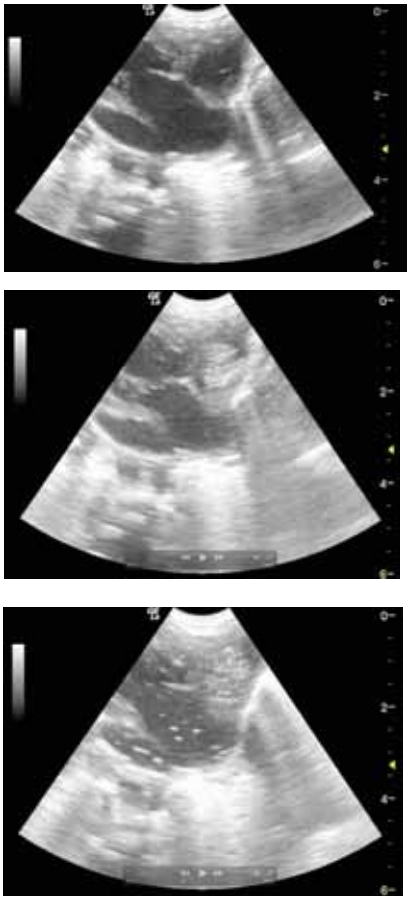

FIGURE 1. Contrast echocardiographic image. A: Filling of the right atrium with bubbles; B: Right atrium with bubbles; C: Filling of the left atrium with bubbles from a venous drain

and the posterior basal segment of the left lower lobe, with the vascular origin in the arteries supplying the same region drained by a dilated (10 mm) common collector. The collector entered the left atrium between the two left pulmonary veins. Two left and two right pulmonary veins also drained into the left atrium. Systemic venous return drained normally through the SVC and inferior vena cava into the right atrium. The left atrium and left ventricle were dilated.

Another small arteriovenous malformation (3 mm diameter) located in the inferior lingular lobe.

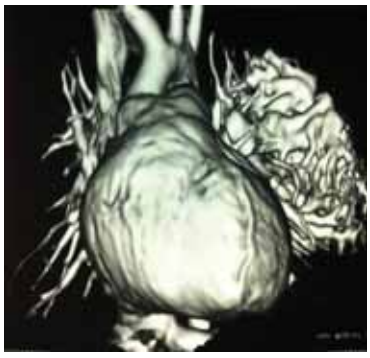

FIGURE 2. Angio CT examination – computerized reconstruction

Pulmonary segmentation was normal, and the rest of the lung appeared to be normal.

Hepatic vascular abnormal image.

She underwent surgery on the 28th day of life. The lungs were exposed through a median sternotomy. The right lung and most of the superior left lung lobe appeared to be normal. The left lower lobe had a dense vascular structure on the costal surface of the lower lobe pedicle with significant dilatation of the lower left pulmonary vein. The left lung was removed. When the left pulmonary artery was ligated, the oxygen saturation increased quickly from 80% to 100%.

Soon after surgery, she developed hypotension and metabolic acidosis. A chest x-ray showed hyperinflation of the right lung and the heart deviated into the left thorax. The patient required increased doses of NO, dopamine, and dobutamine, as well as addition of noradrenaline and milrinone. In the next two days, the pulmonary hypertension improved, and it was possible to decrease the NO and the doses of vasoactive drugs.

On the 14th day after the surgery, it was possible to stop the NO and the milrinone. A few hours later, her pulmonary hypertension worsened again, and she required resumption of NO and initiation of sildenafil.

She also began to have ventricular premature contractions. Echocardiography showed severe right ventricular dysfunction with ejection fraction 25-30%, pulmonary valve insufficiency Gr II, tricuspid insufficiency Gr II, and pulmonary artery dilatation.

She required higher doses of NO, dopamine, dobutamine, and milrinone, and epinephrine was started by continuous infusion.

The clinical status worsened constantly, she developed severe lactic acidosis, which was unresponsive to treatment, and she died on the 48th day of life.

The anatomic pathology examination of the left lung revealed increased vascular territory with only small parts of lung tissue.

DISCUSSION

In neonates, clinical manifestations of the pulmonary arteriovenous malformations from the first days of life are rare. More commonly, these lesions are not diagnosed until the third or fourth decade, when the right-to-left shunt increases and causes symptoms (1). The first PAVM was described in 1975, and since then, about 17 cases have been reported in infants less then one year of age. There is a male predominance of 4:1 in newborns and 1.7:1 in older children; however, in adults, PAVM is more common in females (2). More than half of the PAVMs are located in the lower lobes (3). They are bilateral in 8% to 20% of patients and are multiple in 33% to 50% (4,5).

The pathogenesis of PAVM is not well known. It is hypothesized that it is caused by a defect in terminal arterial loops, which allows dilatation of thin-walled capillary sacs (6) or an incomplete resorption of the vascular septae that separates the arterial and venous plexuses, which normally anastomose during fetal development. Also, multiple small PAVMs can develop as a result of failure of capillary development during fetal development (7).

More then 70% of PAVM cases in adults are associated with hereditary hemorrhagic telangiec- tasia (HHT) know also Osler-Weber-Rendu syndrome (OWRs) (8,9,10). Of OWRs patients with PAVM, 10% also have cerebral arteriovenous malformations (AVMs) and as many as 60% have hepatic AVMs (11). The mortality risk is high for cases of OWRs with PAVM that occur in infancy (12). Although OWRs is a genetic disease (mutations in at least six different genes), the initial diagnosis is based on clinical criteria. The Curacao criteria use the clinical features of epistaxis, multiple telangiectasias, visceral AVM, and a first-degree relative with HHT. A diagnosis of HHT is considered definite if three criteria are present, possible if two criteria are present, and unlikely if fewer than two criteria are present (13).

Our patient had several typical features of OWRs – a large PAVM in the left lower lobe, a second, smaller PAVM in the same lobe, and a small AVMs in the liver. However, our patient met only one of the Curacao criteria – the presence of a visceral AVM- but we had no information about family history of HHT.

A PAVM may be simple, with one or more feeding arteries arising from the same segmental artery, or complex, with multiple feeders from different segmental arteries. Complex PAVMs constitute approximately 10% of the lesions, and a subset of these (approximately 5% of complex PAVMs) has recently been described as diffuse-type. (14)

Depending on the size of the shunt, the typical presentation in neonates is cyanosis, congestive cardiac failure associated with an abnormal chest x-ray showing cardiomegaly, decreased pulmonary vascular markings, and focal pulmonary opacity.

Our patient had a complete type of PAVM and the predominant symptom was severe hypoxemia, requiring ventilation to maintain oxygen saturation.

The suspicion of PAVM was raised by echocardiography and proved by „bubble test". Saline bubbles injected into the superior vena cava appeared first in the right atrium and then, 3 to 5 heart beats later (2-3 seconds), in the left atrium. Left atrial bubbles, due to the intracardiac shunt, would appear immediately after they first appear in the right atrium (15).

Although the PAVM was visible on the chest x-ray and echocardiogram, the final confirmation of the diagnosis came from the angioCT.

PAVMs can sometimes be treated with less invasive approaches, such as coils or balloons. We opted for a more direct surgical approach. Resection of the AVM was considered but was not attempted because of the concerns for the risk of lethal hemorrhage. Left lower lobectomy was also not feasible because of tight adherence of the upper and lower lobes. Instead, the entire left lung was resected.

CONCLUSION

This case illustrates a large pulmonary vascular malformation as a rare cause of hypoxemia and respiratory distress in the newborn. The diagnosis was established using echocardiography, chest x-ray, and confirmed by angioCT. The patient died despite surgical removal of the AVM and the left lung. We think that the patient had Osler-Weber-Rendu syndrome, which, in combination with PAVM, has been reported to have very poor prognosis (16) but also the pneumonectomy had a major effect on worsening the evolution.

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