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Atypical Pulmonary Artery Sling with Diffuse-Type Pulmonary Arteriovenous Fistula

The case of a cyanotic infant with a rare combination of atypical pulmonary artery sling, imperforate anus, absence of the left kidney, interruption of the inferior vena cava, left side hemihypertrophy and diffuse-type pulmonary arteriovenous fistula is described. The clinical features were confusing, because of compounding abnormalities involving the respiratory tract and pulmonary circulation. The diagnostic approach to the etiology of cyanosis is discussed and the embryonic origin of pulmonary artery sling is reviewed.

Key Words : *Pulmonary artery sling; Arteriovenous fistula*

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INTRODUCTION

Classical-type pulmonary artery sling indicates a vascular anomaly in which the anomalous left pulmonary artery rises from the posterior aspect of the right pulmonary artery, passes over the right main stem bronchus near its origin, turns to the left, courses between the trachea and the esophagus and finally reaches the left hilus at a lower level than is normal. Associated anomalies mainly involve the heart or tracheobronchial tree. Extrathoracic anomalies are also rarely associated (1). There have been a few reports of atypical pulmonary artery sling in which both normal and aberrant left pulmonary arteries are present (2, 3, 4).

Most patients with pulmonary artery sling are symptomatic in the first year of life, and nearly half of these, during the newborn period. When present, symptoms are usually those of respiratory obstruction characterized by stridor and wheezing. Obstruction is maximal during expiration and there is no associated dysphagia (1).

We describe a unique case that presented profound cyanosis with atypical pulmonary artery sling, diffuse-type pulmonary arteriovenous fistula, and multiple associated anomalies present.

CASE REPORT

An eight-month-old male infant presented with progressive respiratory distress and profound cyanosis. He was the first baby of a healthy unrelated couple who had an uncomplicated pregnancy. There was no maternal history of medication, illness, smoking or alcohol consumption during pregnancy. He weighed 2.98 kg at birth and gestation period was 39 weeks. Imperforate anus was noted after birth and colostomy was performed at three days of age. At five months, he developed severe respiratory distress caused by an upper respiratory infection and was treated in a hospital for three weeks under the impression of bronchiolitis. Blood gas analysis during admission showed a pH of 7.2-7.4, PCO_2 of 40-50 torr, PO_2 of 133-439 torr, and oxygen saturation of 97-99.8%. The second episode of severe respiratory distress following upper respiratory infection developed two months later and cyanosis was then first noted. Cyanosis and respiratory distress subsequently progressed with intermittent episodic aggravation.

On admission, the patient looked ill and dyspneic; moderate cyanosis was found throughout the whole body, including on the lips and tongue. His face did not appear dysmorphic but hemihypertrophy was noted on the left side. Vital signs were blood pressure 80/40 mmHg, heart rate 133/min, respiration rate 56/min, weight 7.6 kg (3-10 percentile) and height 78 cm (90-97 percentile). Chest expansion was symmetric, with suprasternal and subcostal retractions. Breathing sound was coarse without rales, stridor or wheezing, heart rhythm was regular and murmur was not heard. The abdomen was slightly tense, liver and

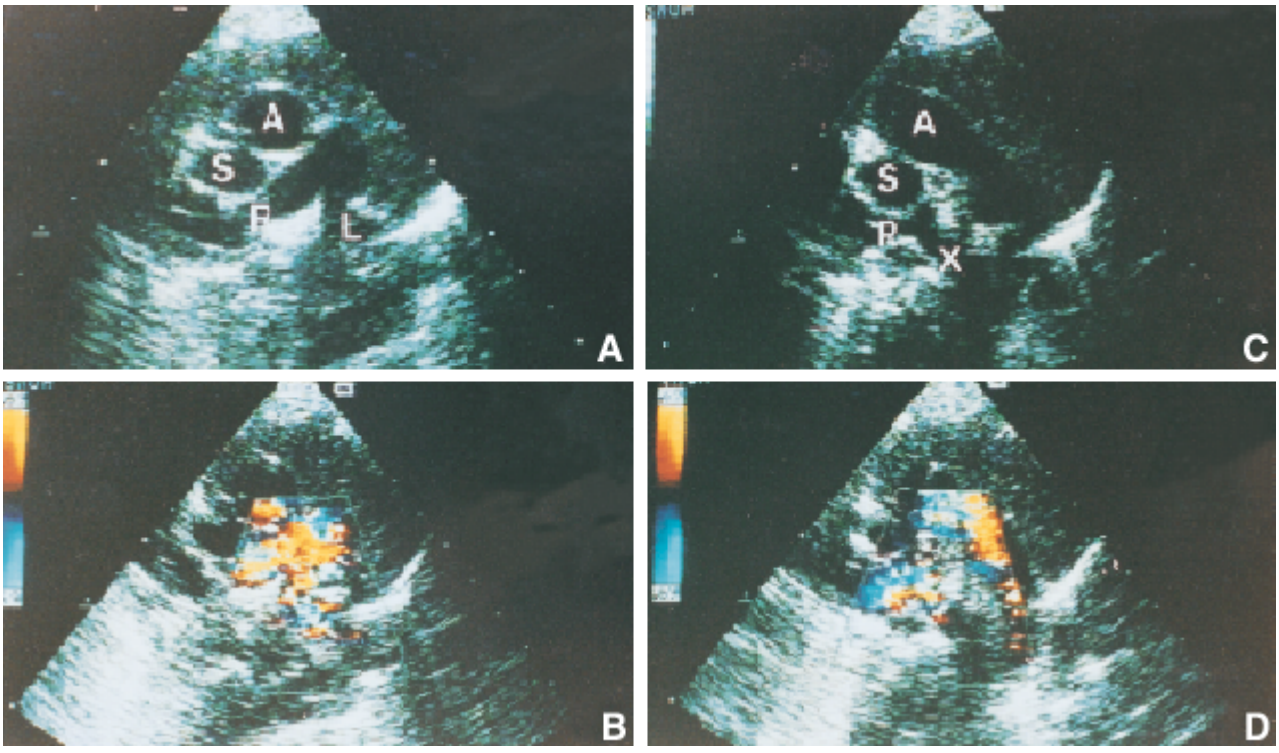

Fig. 1. Echocardiogram from the parasternal short axis view demonstrates normal and anomalous left pulmonary arteries arising from the right pulmonary artery. A. Right and left pulmonary arteries originate normally. B. Color Doppler demonstrates normal flow in both pulmonary arteries. C. Aberrant left pulmonary artery rises from the right pulmonary artery distal to the normal bifurcation site of the left pulmonary artery. D. Color Doppler demonstrates flow through aberrant left pulmonary that courses behind the trachea. A, aorta; S, superior vena cava; R, right pulmonary artery; L, left pulmonary artery; X, aberrant left pulmonary artery.

spleen were not palpable. An operation scar and colostomy were seen on the right lower quadrant of the abdomen. There was mild clubbing of fingers and toes. Both femoral pulses were clearly palpated and symmetrical.

Complete blood count and liver function test revealed no significant abnormalities and chromosomal study was normal. A chest radiograph showed a normal-sized heart and pulmonary vascular markings were slightly increased. Both lungs were hyperexpanded. High penetration film showed a narrowing of the lower trachea, long segment narrowing of the right bronchus and short segment narrowing of the left bronchus; surgery thus appeared very difficult. An electrocardiograph showed a P-wave axis of -45 degrees and a QRS axis of -82 degrees. There was no evidence of ventricular hypertrophy. Arterial blood gas analysis in room air showed a pH of 7.33, PCO_2 of 43 torr, PO_2 of 32 torr, and oxygen saturation of 56%. After high flow oxygen administration, PO_2 increased to 43 torr and oxygen saturation to 72%. PCO_2 remained the same, 39 torr. Methemoglobin was 0.4 g/dL.

Two-dimensional echocardiography showed a normally connected heart without intracardiac abnormalities but with interruption of the inferior vena cava. At first sight, the main pulmonary artery and its branches appeared normal. Closer observation, however, revealed another aberrant left pulmonary artery rising from the right pulmonary artery, and coursing behind a highly reflective air containing structure, possibly the trachea (Fig. 1). Color-flow mapping confirmed normal and aberrant left pulmonary arteries. Contrast echocardiography, in which indocyanine green was infused into the peripheral vein showed opacification in the left atrium after four or five cardiac cycles. Abdominal ultrasound examination showed that the left kidney and inferior vena cava were absent. The right kidney was normal and visceral position was reported to be normal.

Although risky, it was decided to undertake cardiac catheterization, because the pulmonary arterial anatomy was very unusual and cyanosis was not explained solely by airway compression. Cardiac catheterization was performed under sedation. When venous and arterial access were obtained, the patient's respiratory symptoms became aggravated. Catheterization was minimal, consisting of blood sampling and pressure measurement of the right and left side of the heart. Immediately after measurement, oxygen was administered and external respiratory support was provided, and his condition soon stabilized. Angiography was then performed. Pulmonary artery (42/6 (24) mm Hg) and right

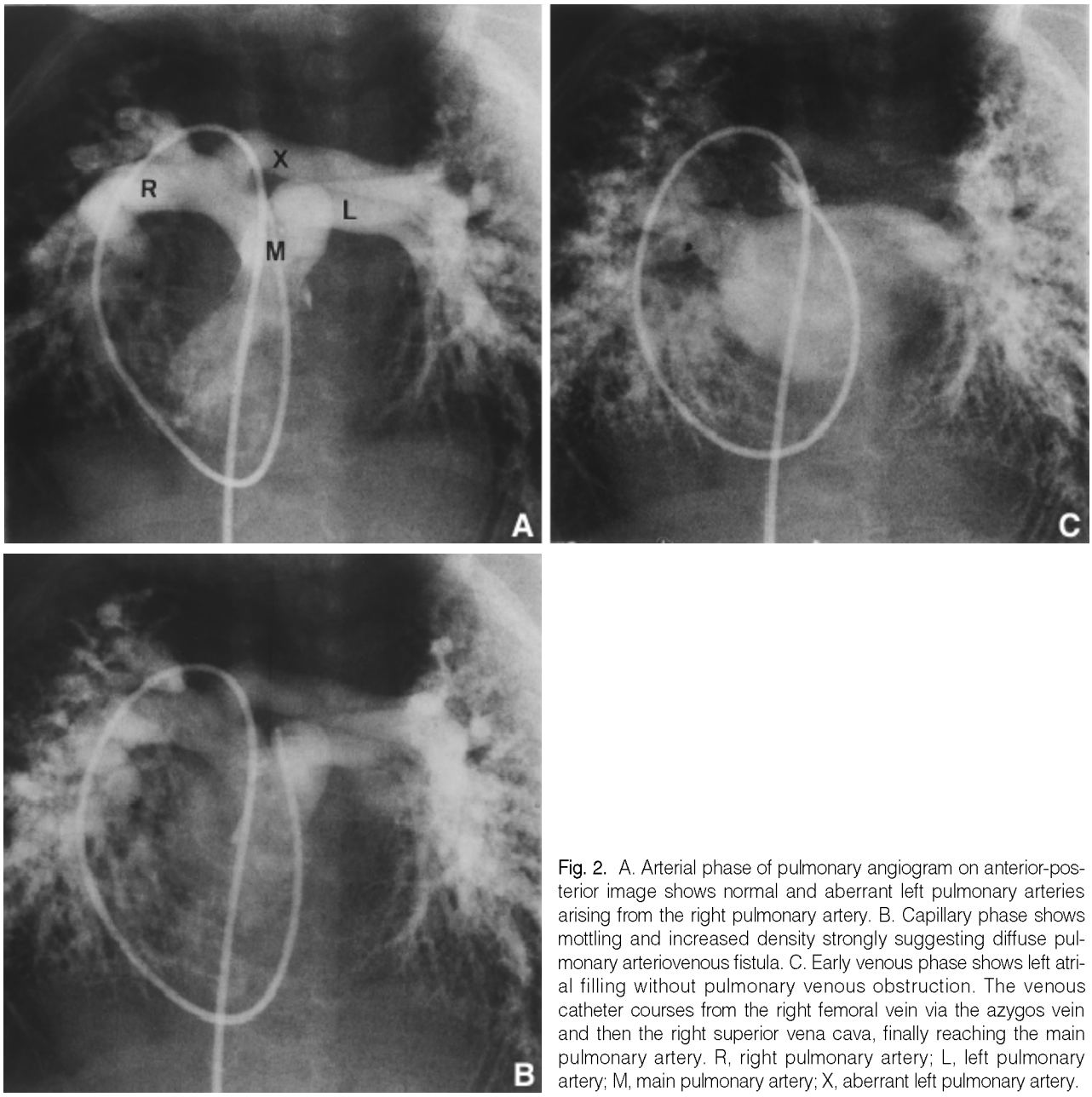

Fig. 2. A. Arterial phase of pulmonary angiogram on anterior-posterior image shows normal and aberrant left pulmonary arteries arising from the right pulmonary artery. B. Capillary phase shows mottling and increased density strongly suggesting diffuse pulmonary arteriovenous fistula. C. Early venous phase shows left atrial filling without pulmonary venous obstruction. The venous catheter courses from the right femoral vein via the azygos vein and then the right superior vena cava, finally reaching the main pulmonary artery. R, right pulmonary artery; L, left pulmonary artery; M, main pulmonary artery; X, aberrant left pulmonary artery.

ventricle pressure (48/6 mmHg) were moderately elevated, though left ventricle and aorta pressure were normal. Oxygen saturation of the aorta was 21.5% and high-flow oxygen administration under a hood for about 30 minutes increased it to a maximum of 78%. Interruption of the inferior vena cava and an atypical pulmonary artery sling was confirmed by angiography (Fig. 2). The normal left pulmonary artery arose from the main pulmonary artery, coursed over the left bronchus and supplied the left upper and lower lung fields. The aberrant left pulmonary artery arose in the middle of the right pulmonary artery, coursed behind the trachea and supplied the left middle lung fields. Because of the patient's poor condition and difficulty in manipulating the catheter, each left pulmonary artery was not selectively injected. The capillary phase showed generalized fine mottling of the whole lung field and early appearance of the pulmonary vein, strongly suggesting diffuse-type pulmonary arteriovenous fistula. Aortography showed an abnormally positioned abdominal aorta on the right side.

The patient's condition deteriorated progressively but his parents declined further diagnostic tests and aggressive management. He died at eight months of age and autopsy was

refused.

DISCUSSION

Pulmonary artery sling has been reported to be associated with many congenital anomalies, not only of the cardiovascular and tracheobronchial system but also of other organs. Associated extrathoracic anomalies reported to date include imperforate anus, absent gallbladder, hemivertebra-agenesia left kidney, ureterovesical obstruction, diaphragmatic hernia, trisomy 21 and so on (1). Associated congenital anomalies are important in determining both managerial approaches and ultimate prognosis.

The case described in this paper showed multiple anomalies, some of which have been reported as associated. These include imperforate anus and absence of the left kidney. However, the association of other anomalies such as interruption of the inferior vena cava, left side hemihypertrophy and pulmonary arteriovenous fistula with pulmonary sling has not been previously reported. Interruption of the inferior vena cava is commonly associated with left atrial isomerism. However, there was no other evidence of visceral heterotaxy or intracardiac defect. Klippel-Trenaunay-Weber syndrome, which consists of asymmetric limb hypertrophy, hemangiomata, varicose vein and interruption of the inferior vena cava has never been associated with pulmonary arteriovenous fistula (5). Because of absence of the facial and limb abnormalities, Potter syndrome was excluded (6).

Since the first report of pulmonary artery sling in 1897, the embryology of this lesion has not been firmly established. It has been proposed that pulmonary artery sling arises when the left lung bud fails to connect with the left sixth arch and, instead, forms an anastomosis to the right sixth arch dorsal to the developing lung bud. Jue et al. (7) proposed that such a connection might occur ventral to the developing lung bud rather than dorsal to it. As illustrated in Fig. 3, the left pulmonary artery rises aberrantly from the right pulmonary artery and can take a ventral course to the trachea (8). Moreover, both normal and aberrant left pulmonary artery can coexist. In these cases, aberrant left pulmonary artery take a ventral or dorsal course (2, 3, 4). The embryologic hypothesis to account for aberrant left pulmonary artery seems to be, therefore, that part or the whole of the left postbranchial plexus connects ventrally or dorsally with the right pulmonary artery. We still do not know, however, why dorsal connection occurs much more frequently and why the aberrant origin of the right pulmonary artery has not been reported. Experimental and molecular studies of pulmonary arterial development seem necessary.

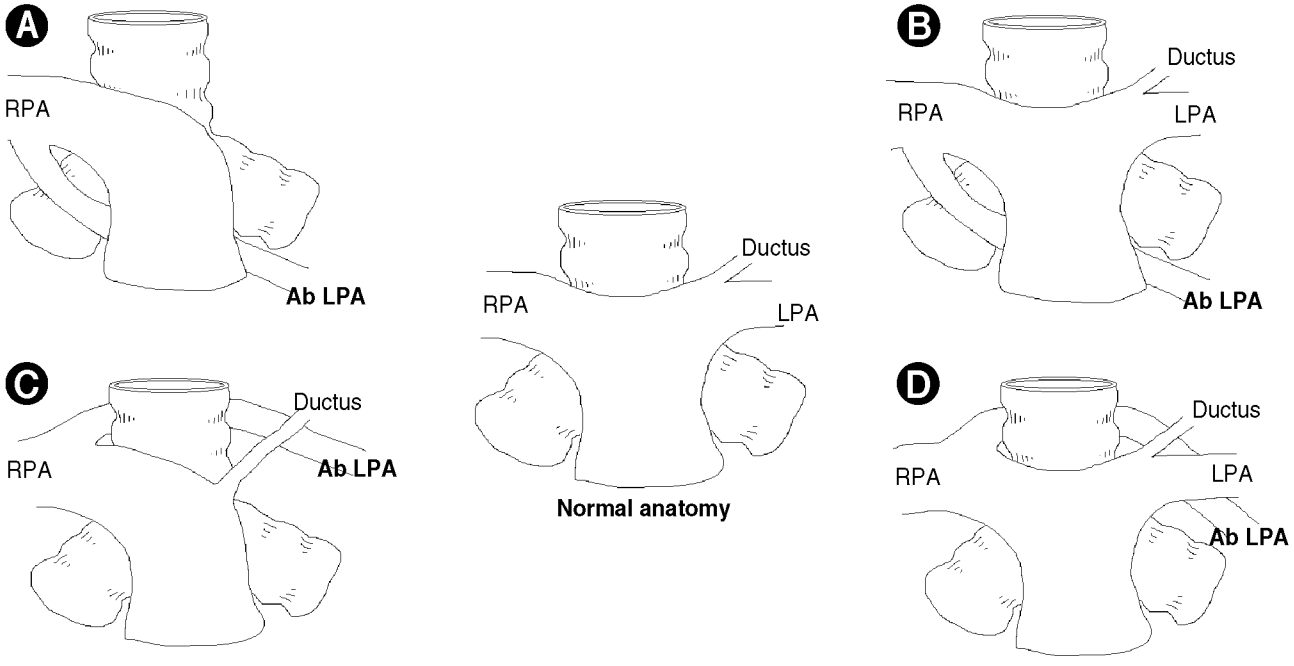

Fig. 3. Four varieties of anomalous pulmonary artery. Normal anatomy is shown in the center. A. LPA arising ventrally from the RPA passing anterior to the trachea (Moreno et al., Ped Cardiol (1991)). B. Partial sling with normal LPA and aberrant LPA arising from the RPA passing anterior to the trachea (Erickson et al., Ped Cardiol (1996)). C. Usual sling development with anomalous LPA arising dorsally from the RPA passing posterior to the trachea. D. Partial sling with normal LPA and aberrant LPA arising from the RPA and passing posterior to the trachea (Gikonyo et al., Ped Cardiol (1989); Bamman et al., Chest (1977); our case). RPA, LPA, right, and left pulmonary arteries; Ab LPA, aberrant left pulmonary artery; Ductus, ductus arteriosus.

Pulmonary artery sling is usually suspected when clinical symptoms of respiratory obstruction are associated with anterior indentation of the esophagus on barium swallowing. Various imaging techniques have been used to confirm diagnosis. Echocardiography and other non-invasive studies proved to be extremely useful for the diagnosis of pulmonary artery sling (9, 10). In typical cases, the left pulmonary artery does not arise normally, but distally from the right pulmonary artery. In atypical cases, however, normal left pulmonary artery does exist and it is therefore important to be aware that its existence does not preclude aberrant origin of the left pulmonary artery. Because of the highly reflective air-column, it is difficult to assess an airway by echocardiography, but the very nature of its characteristics enables its localization in relation to vascular structures.

A pulmonary arteriovenous fistula has been defined as an abnormal connection between the pulmonary arterial and venous system that bypasses the pulmonary capillary bed. It may be a solitary lesion, multiple lesions, or a form of diffuse telangiectasia (11, 12). Pulmonary arteriovenous fistula is very rare and most are congenital either as a component of hereditary hemorrhagic telangiectasia or in an isolated form. A few cases may be acquired. They can occur after trauma, develop after Glenn shunt or Fontan procedure, or complicate liver disease. The most common symptom in childhood is cyanosis, and hemoptysis may occur. Compared with cyanosis of respiratory origin, that of pulmonary arteriovenous fistula is not much affected by oxygen inhalation.

The patient described in this paper had suffered from progressive cyanosis and respiratory distress since five months of age. Echocardiography showed no intracardiac defect but an atypical form of pulmonary artery sling, which seemed to be responsible for his respiratory symptoms. Severe cyanosis which showed limited response to the administration of high-flow oxygen was not explained solely by airway obstruction. Moreover, blood gas analysis showed a normal PCO_2 level. Another unusual finding showed dilated pulmonary veins on two-dimensional echocardiography, which suggested increased pulmonary blood flow and the possibility of pulmonary arteriovenous fistula. Contrast echocardiography and pulmonary angiography confirmed the presence of the generalized telangiectatic form of pulmonary arteriovenous fistula. Therefore we concluded that in this case, cyanosis was mainly due to pulmonary arteriovenous fistula and symptoms of respiratory obstruction due to pulmonary artery sling. It is not clear whether pulmonary arteriovenous fistula is congenital or acquired. The fact that cyanosis was not evident until seven months of age may suggest that pulmonary arteriovenous fistula is acquired. The possibility that it is congenital is not excluded, however; if the congenital form is mild, it may not present during the newborn period.

A case in which there is a very unusual combination of anomalies has been described. Although autopsy was not performed, important anomalies were properly assessed and characterized by clinical means and we do not think that there were any major other organ anomalies that were missed or undiagnosed. Recent refinement of many imaging techniques makes it possible to confirm cardiovascular anomalies, which in the past required invasive studies. Among many non-invasive techniques, echocardiography has proved to be extremely useful. In this case, an atypical form of pulmonary sling was reliably diagnosed by echocardiography. This case also illustrates that pulmonary arteriovenous fistula can progress rapidly and be fatal, and also that normal origin of the left pulmonary artery does not preclude an atypical form of pulmonary artery sling.

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