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INTERRUPTION OF THE DISTAL LEFT PULMONARY ARTERY WITH PULMONARY ARTERIOVENOUS FISTULAS AND ATRIAL SEPTAL DEFECT*

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A seven-year-old boy with interrupted left pulmonary artery, arteriovenous fistulas and atrial septal defect is presented. The anomalies were identified by echocardiography and angiography. On pulmonary artery angiograms, left pulmonary artery discontinuity (as though the distal segment had been interrupted), diffuse granulary opacification of right lung fields, and early visibility of the left atrium were observed. *Key words: pulmonary artery anomaly, interruption, arteriovenous fistula.*

Herein we present a seven-year-old boy who had interruption of the distal half of the left pulmonary artery, diffuse pulmonary arteriovenous fistulas, and atrial septal defect. Although anomalies of the pulmonary artery, such as pulmonary artery stenosis at different levels and unilateral absence of a pulmonary artery, have been well described[1-3], our case had a different kind of pulmonary artery anomaly, which can be described as an "interruption". To our knowledge, this is the first reported case in literature of this anomaly in association with pulmonary arteriovenous fistulas and atrial septal defect.

Case Report

A seven-year-old boy was admitted to our hospital with dyspnea, cyanosis and frequent lower respiratory tract infections of five years duration. On physical examination, diffuse cyanosis of a moderate degree, clubbing, and growth retardation were detected. The precordium was prominent with hyperactive right ventricular impulse. On auscultation, a mild ejection murmur was heard at the upper left sternal border, and the second heart sound was increased. Hemoglobin was 16 g/dl, and hematocrit was 54 percent. Chest x-ray showed a moderate degree of cardiomegaly, diffuse reticular infiltration and small rounded opacities in the right lung fields, and a prominent main pulmonary artery segment.

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Electrocardiogram revealed right axis deviation and right ventricular hypertrophy. Two-dimensional echocardiography showed a secundum atrial septal defect and a mildly enlarged main pulmonary artery. Contrast echocardiography demonstrated rapid visibility of the left atrium, and also an atrial septal defect. On M-mode echocardiogram, the right ventricular PEP/VET ratio (pre-ejection period/ventricular ejection time) was greater than 0.3, which was a sign of pulmonary hypertensilon. Tricuspid regurgitation, with a velocity of 2.90 m/sec, was detected by Doppler echocardiography.

Right heart catheterization was performed through the femoral venous route. Systemic arterial oxygen saturation was 78 percent, which was detected in samples obtained from the pulmonary veins. Pressures were the same and increased both in the right and left pulmonary arteries with systolic 60 and diastolic 20 mm Hg. Right and left ventricular systolic pressures were 60 and 80 mm Hg, respectively.

Selective pulmonary angiograms showed multiple small, rounded opacifications disseminated on the right lung fields (Fig. 1). Early opacification of the left atrium was also seen. The angiography of the left pulmonary artery demonstrated an absence of its normal distal portion, as though it were interrupted with a sharp edge. Thin and collateral-like vessels were disseminating throughout the left lung fields from the interrupted left pulmonary artery (Fig. 2). A small atrial septal defect and left-to-right shunt were detected by left atrial angiography and oximetric data. The pulmonary to systemic flow ratio was 1.5/1, and pulmonary vascular resistance was 3 Wood units/m^2.

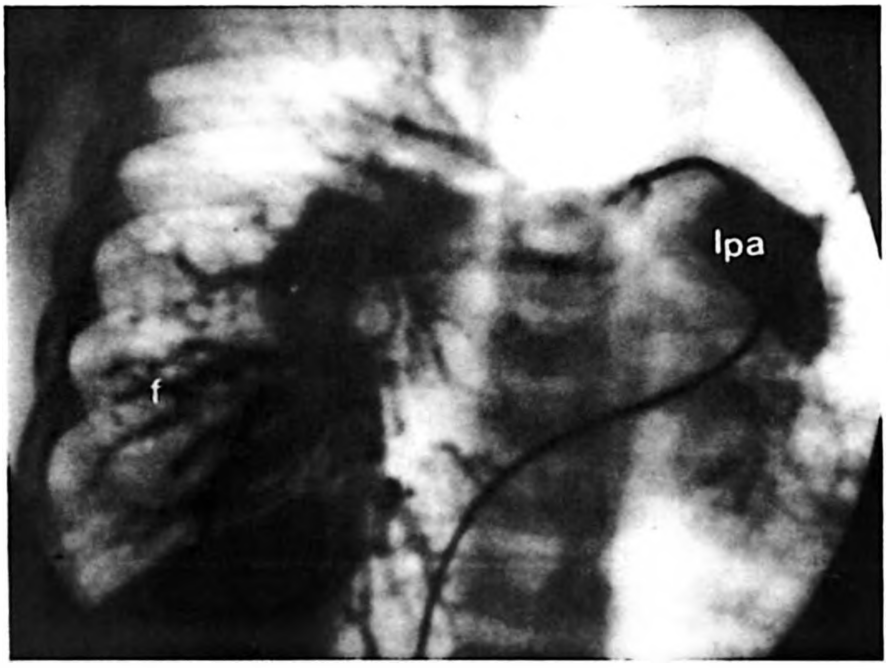

Fig. 1: Pulmonary arteriography in the anteroposterior projection. Multiple opacifications on the right lung fields due to arteriovenous fistulas and discontinuity of the left pulmonary artery are shown. f: arteriovenous fistulas, lpa: left pulmonary artery.

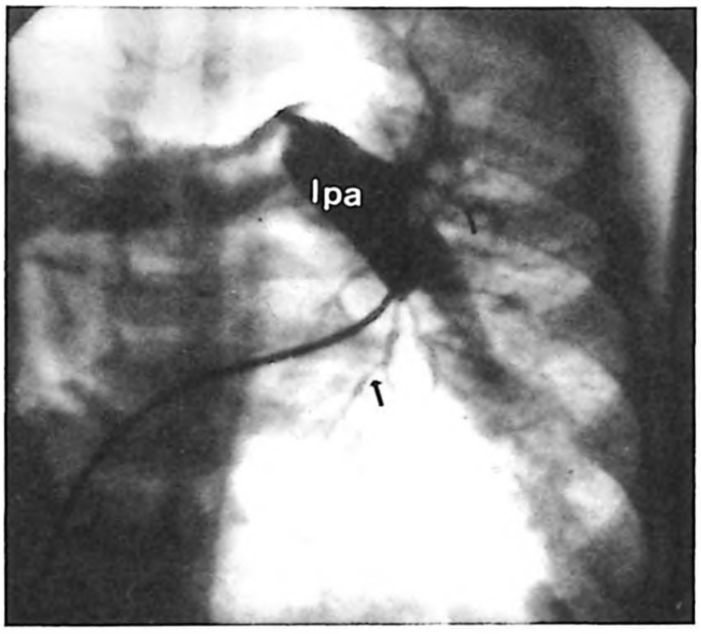

Fig. 2: The left pulmonary artery angiogram in the anteroposterior projection demonstrates the interrupted left pulmonary artery with thin collateral-like vessels (arrows) disseminating throughout the left lung fields. lpa: left pulmonary artery.

Discussion

Unilateral absence of one pulmonary artery, first described in 1868, is an uncommon congenital defect. Other cardiac defects, such as atrial septal defect, patent ductus arteriosus, and truncus arteriosus, have been described with this anomaly[5]. In our patient, the left pulmonary artery was not completely absent: the proximal half of the vessel was normally developed, with even some degree of enlargement, despite the absence of the distal part. Presbitero et al.[2] identified the previously mentioned absent pulmonary arteries on surgical examination and described than as interrupted pulmonary arteries. This also correlated with the pathologic findings of McKim and Wiglesworth[6]. Thus, we described this unilateral left pulmonary artery discontinutiy (or unilateral non-confluent left pulmonary artery with absence of distal part) as interrupted left pulmonary artery[7,8]. In our patient, diffuse pulmonary arteriovenous fistulas on the right lung and a secundum atrial septal defect were also associated with this pulmonary artery anomaly. Pulmonary hypertension was present, but its degree of severity was not correlated with the size of the atrial septal defect.

Pulmonary arteriovenous fistulas are almost always congenital in origin. The disease may also be acquired as a result of many conditions, such as schistosomiasis, juvenile cirrhosis, thyroid carcinoma, and trauma[9]. The majority of patients with pulmonary arteriovenous fistulas have an associated Rendu-Osler-Weber syndrome. Absence of the right lower lobe of the lung and

congenital heart diseases have been described in association with fistulas[10]. In this rare case of pulmonary arteriovenous fistulas with an absence of the distal left pulmonary artery, fistulas may either be congenital in origin, where a combination of these two disorders may be coincidental or, less likely, may be acquired due to hypoperfusion of the left lung.

It is well known that surgical resection is the preferred treatment for arteriovenous fistulas which are not too disseminated. Balloon embolization is an alternative therapy for patients at high risk surgically or whose lesions are too diffuse to resect[11]. New techniques have also been described for surgical repair of unilateral non-confluent pulmonary arteries[12]. Our patient was considered inoperable because of his pulmonary hypertension and diffuse arteriovenous malformations. Congenital heart diseases have been described in association with pulmonary fistulas. However, to our knowledge, this is the first case of a different form of pulmonary artery anomaly (such as an interruption of the left pulmonary artery) with pulmonary arteriovenous fistulas on the other lung, atrial septal defect, and pulmonary hypertension.

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