DIRECT COMMUNICATION BETWEEN RIGHT PULMONARY ARTERY AND LEFT ATRIUM

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RESUMEN

COMUNICACIÓN DIRECTA ENTRE LA RAMA DERECHA DE LA ARTERIA PULMONAR Y LA AURÍCULA IZQUIERDA

Presentamos un caso de comunicación directa entre la arteria pulmonar derecha y la aurícula izquierda. El diagnóstico se hizo antes de la operación. La ligadura quirúrgica de la fístula, resolvió la cianosis de la enferma. El angiocardiograma selectivo en la arteria pulmonar derecha, realizado 4 meses después, descartó cortocircuito residual. Esta malformación muy rara debe considerarse en presencia de cianosis inexplicable. Es éste el caso número 50 en la literatura.

SUMMARY

A case of direct communication between right pulmonary artery and left atrium is reported. The diagnosis was made before surgical correction. A surgical ligation of the fistula resolved the cyanosis of the patient. Selective angiocardiogram of the right pulmonary artery 4 months after surgery revealed no residual shunt. This very rare malformation should be considered in the clinical setting of unexplained cyanosis. This is the number 50 case reported in the literature.

RESUME

COMMUNICATION DIRECTE ENTRE LA BRANCHE DROITE DE L'ARTERE PULMONAIRE ET L'OREILLETTE GAUCHE

On présente un cas de communication directe entre l'artère pulmonaire droite et l'oreillette gauche. Le diagnostic a été établi avant l'intervention chirurgicale. La ligature de la fistule élimina la cyanose de la malade. L'angiocardiogramme sélectif dans l'artère pulmonaire droite, effectué 4 mois après le traitement chirurgical, écarta la possibilité d'un shunt résiduel. Cette malformation, assez rare, doit être envisagée en présence d'une cyanose inexplicable. Le cas, qu'on relate ici, est le No. 50 de la littérature.

Palabras clave: Fístula pulmonar congénita. Comunicación directa entre la arteria pulmonar y la aurícula izquierda. Enfermedad congénita cardíaca.

Key words: Pulmonary congenital fistula. Direct communication between right pulmonary artery and left atrium. Congenital heart disease.

INTRODUCTION

Direct communication between the right pulmonary artery and the left pulmonary artery is an extremely rare anomaly, up to date 50 cases have been published, including our case.[1]

The aim of this report is to describe a case report with her surgical correction.

Case report

A 6 year old girl was admitted to our hospital. Clinical history revealed cyanosis and dyspnea on effort since the age of 5. Physical examination confirmed central cyanosis and clubbing, arterial pressure 100/70 mmHg, cardiac rate 72 per minute, respiratory rate 24. Precordial and cardiac sounds were

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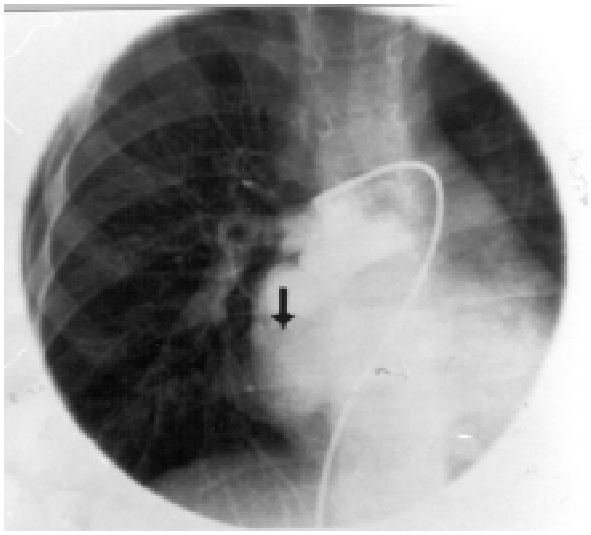

FIG. 1A: A selective angiocardiogram in the right pulmonary artery showed a large saccular communication *(arrow)* between the right pulmonary artery and the left atrium, (frontal projection).

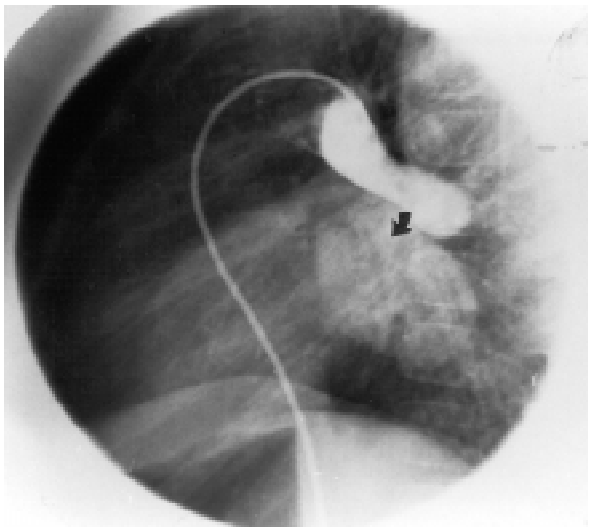

FIG. 1B: Lateral view.

normal. At the right side of the chest, a systolic soft murmur grade I/IV was audible. Chest ray and ECG, as well as segmental analysis by bidimensional echocardiography, were normal; in addition, only slight left chambers enlargement was noticed. Cardiac catheterization showed mean pressure in the right atrium of 3, right ventricle 14/3, pulmonary trunk 14/6/9 mmHg with normal oxygen content in the right side of the heart; however, a blood desaturation was detected from the pulmonary vein (93%) to the left atrium (82%). Oxygen content remained low in the left ventricle and aorta. Mean pressure in the left atrium was 4, left ventricle 100/4 and aorta 110/75/86 mmHg. Angiocardiography performed in the right pulmonary artery revealed a saccular communication between this vessel and the left atrium *(Figure 1)*. Distal injections in both pulmonary arteries allowed to visualize normal progression of the contrast material in the lungs. The patient underwent surgery, by sternotomy the right hilium was dissected, the abnormal fistula identified and ligated. An increase in the arterial saturation was detected immediately. She recovered uneventful during the four days after the operation, at the fifth day she developed unexpected generalized convulsions, followed by hemiparesia. An echocardiogram showed no residual thrombus.

Craneal tomographic scans disclosed occipital and lacunar infarctions. Four months later, the patient is doing well with no residual sequela. A control selective angiocardiogram in the right pulmonary artery revealed no residual shunt *(Figure 2)*.

DISCUSSION

Direct communication between the right pulmonary artery and the left atrium should be considered when other usual causes of cyanosis have been excluded. The bidimensional echo study should revealed the diagnosis taking in mind this anomaly. This malformation is considered as a distinct entity, quite apart from the well known pulmonary arteriovenous fistula.[2] All reported cases have shown cyanosis, from birth to late adulthood.[1-4]

The following complications have been described: systemic embolism, cerebral abscess.[5-7] Our patient presented systemic embolism, probably related to a residual thrombous in the left atrium. Associated lesions reported are hemangioma of the forehead,[8] stenosis of the left pulmonary artery,[5] varicose veins of the legs, and pulmonary valve regurgitation.[2]

Embryologically this malformation may result from a fistula, during early embryonic development, between the pulmonary artery and the main primitive pulmonary veins.

Later the large pulmonary vein becomes incorporated into the wall of the left atrium during atrial enlargement.[5] This is supported by the fact that in

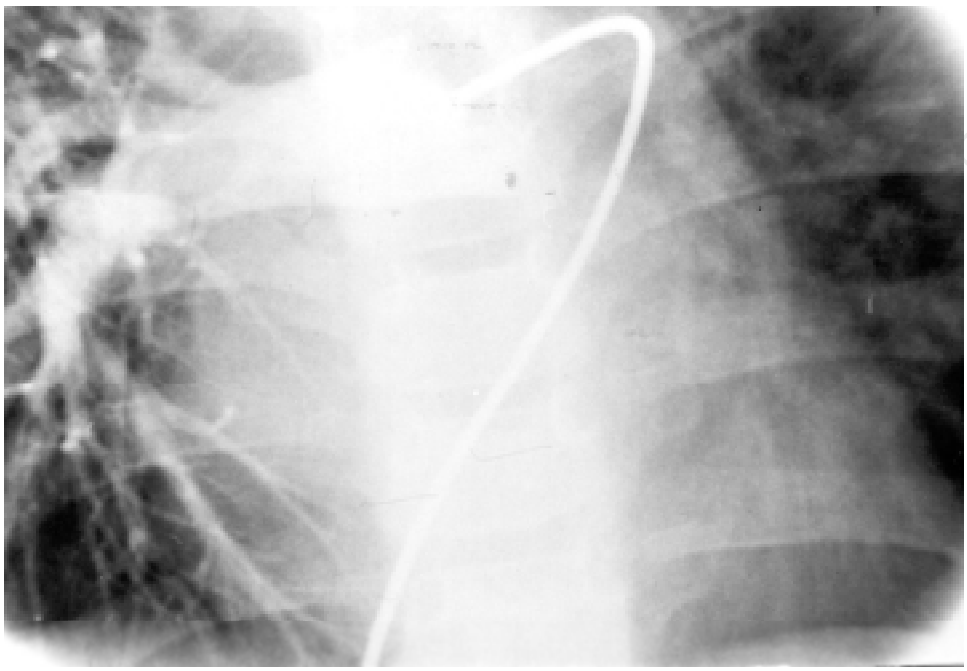

FIG. 2A: Control angiocardiogram 4 months later, revealed no residual shunt, (frontal projection).

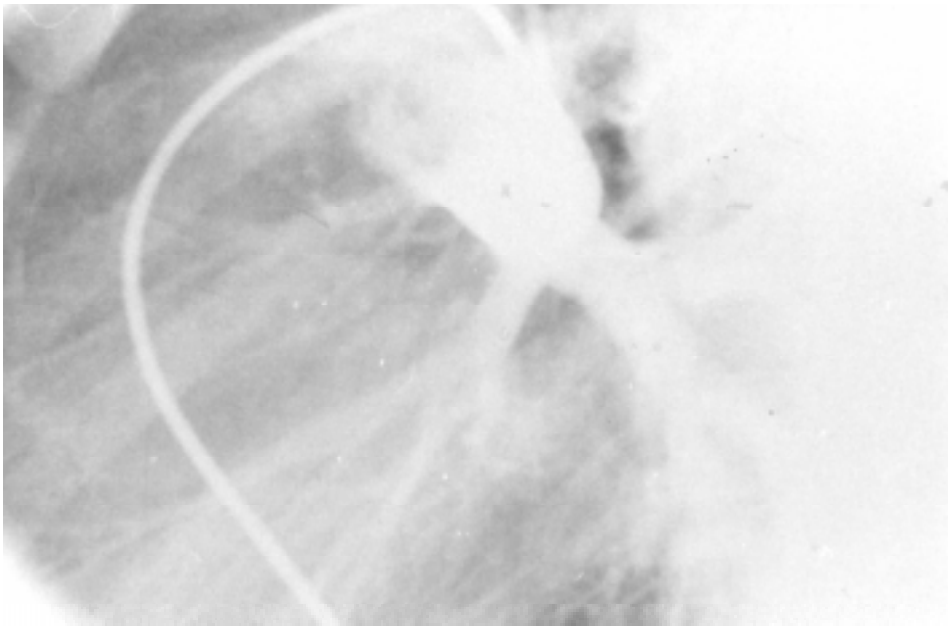

FIG. 2B: Lateral view.

some cases the pulmonary veins and the right pulmonary artery drain into a saccular dilatation before entering the left atrium.[8] In our case, the fistula was saccular in shape, however, the pulmonary veins connected properly with the left atrium. Surgical ligation of the fistula with or without cardiopulmonary bypass has been the appropriate treatment.[1-4]

In conclusion, in patients with late onset cyanosis and normal heart auscultation, this anomaly should be considered once the more usual diagnose have been laid aside. This is the first case reported in Mexico.

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