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Case Report

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Pulmonary artery dissection. Case report

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ARTICLE INFORMATION

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Acronyms
ASD: atrial septal defect
PAD: pulmonary artery dissection
RA: right atrium
RV: right ventricle

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Spanish - English

ABSTRACT
The dissection of the pulmonary artery is an extremely rare disease that is related to the presence of severe pulmonary hypertension of long evolution, in the course of congenital heart diseases or primary pulmonary hypertension. This is the case of a woman with a history of ostium secundum type atrial septal defect that refused to undergo surgical treatment and presented suprasystemic pulmonary hypertension secondary to Eisenmenger's syndrome, in the course of its natural evolution, and comes to our hospital complaining of chest pain and dyspnea at rest, and where pulmonary artery dissection was diagnosed.
Key words: Pulmonary artery dissection, Eisenmeger's syndrome, congenital cardiac diseases

Disección de arteria pulmonar. A propósito de un caso

RESUMEN
La disección de la arteria pulmonar es una enfermedad extremadamente rara que se relaciona con la presencia de hipertensión pulmonar grave de larga evolución, en el curso de cardiopatías congénitas o de hipertensión pulmonar primaria. Se presenta el caso de una mujer, con antecedentes de comunicación interauricular tipo *ostium secundum* que se negó a recibir tratamiento quirúrgico y presentó hipertensión pulmonar suprasistémica que desarrolló un síndrome de Eisenmenger en el curso de su evolución natural, y acude a nuestro centro por dolor precordial y disnea en reposo donde se diagnostica disección de la arteria pulmonar.
Palabras clave: Disección de arteria pulmonar, Síndrome de Eisenmenger, Cardiopatías congénitas

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INTRODUCTION

Atrial septal defect (ASD) is the most common acyanotic congenital heart disease in adults,and is present in 40 % of adults with congenital heart disease[1].

If this disease is not diagnosed and corrected at an early age, it can condi-

tion the development of pulmonary hypertension in adulthood, with the consequent dilation of the pulmonary tree[2] and the possibility of developing pulmonary artery (PA) dissection.

According to the literature reviewed, the dissection usually occurs at the site where the pulmonary vascular wall becomes very labile in relation to changes in existing pulmonary pressures.

According to Senbaklavaci et al.[3], the first clinical case was presented post-mortem in 1862 by Walshe. Since then, there have been few reports of patients diagnosed by invasive techniques such as pulmonary angiography, or post- mortem studies in 50 % of cases, which is due to their frequent occurrence as cardiogenic shock or sudden death, although there are reports of patients whose symptoms are limited to chest pain or dyspnea at rest and who have been diagnosed during life[4].

This article describes, for the first time in Cuba, the diagnosis *in vivo* of a patient with dissection of PA.

CASE REPORT

52-year-old white female, with a history of uncorrected ostium secundum type ASD and pulmonary hypertension since 1983, developing Eisenmenger syndrome in the course of its natural evolution who attended our hospital with an episode of chest pain accompanied by dyspnea at rest associated with cough and hemoptysis, for which she was hospitalized.

On physical examination distressing facies, perioral cyanosis (**Figure 1A**), hippocratic fingers (**Figure 1B**), pectus carinatum and pedal edema were found. At the level of the respiratory system an overall decrease of breath sounds and crackles until the middle third of both hemithorax was noticed. On examination of the cardiovascular system, increased amplitude arterial pulse, increased jugular venous pressure, cardiac apex shifted to the fourth left intercostal space and palpable right ventricle (RV) were found. On auscultation a first normal noise was found, the second was accentuated at the expense of pulmonary component, with constant split, presence of fourth right noise and early diastolic murmur IV/VI (Graham -Steel) audible in pulmonary area that radiated to the left sternal edge. In addition, in tricuspid focus a holosystolic murmur that increased with inspiration, intensity III/VI, radiating to the xiphoid appendix, right sternal border and left anterior axillary line was auscultated. In the abdomen hepatomegaly was palpated with left lobe predominance, which exceeded in 4 cm the costal margin, and hepatojugular reflux maneuver was positive.

Hemo-biochemical parameters were within normal limits.

The surface electrocardiogram showed an increase in voltage of the P wave, pattern of right bundle branch block and right ventricular hypertrophy with signs of systolic overload.

The chest radiograph (posterior- anterior view) revealed a great cardiomegaly with prevalence of RV with huge dilated right atrium (RA) and aneurysmal dilation of PA (**Figure 1C**). Furthermore, the absence of vasculature to the periphery of both lungs (outer

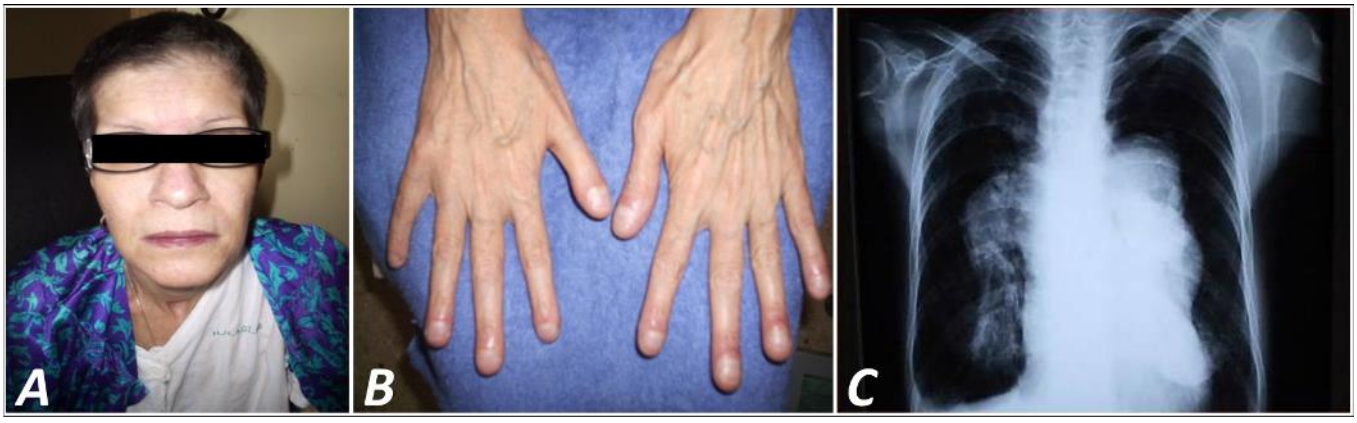

Figure 1. A. Presence of perioral cyanosis. **B.** Hippocratic or "drumstick" fingers. **C.** Chest X-Ray. Note the huge expansion of the middle arch and pulmonary arterial branches, and the absence of peripheral pulmonary vasculature.

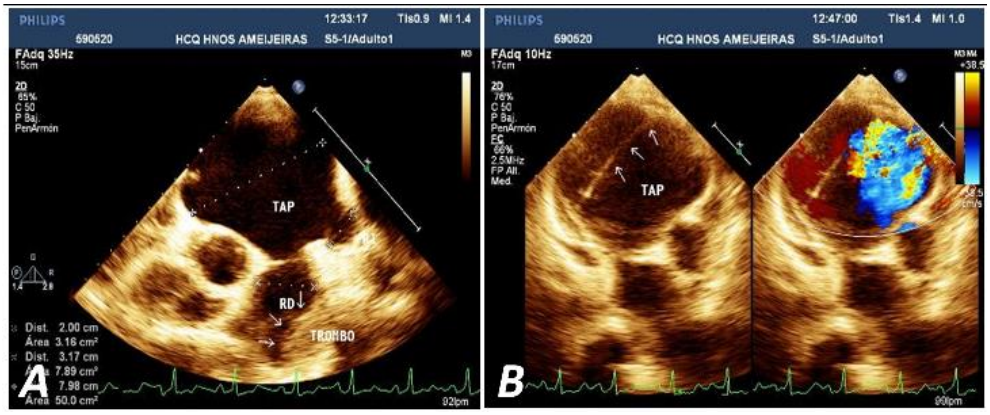

Figure 2. A. Aneurysmal dilation of PA and its branches, with the presence of thrombus in the right branch (RB). **B.** Intimal flap (arrows) with slowing flow and communication in the false lumen.

third) typical of long-standing hypertension, what some people call "lung fields".

On echocardiography, the great growth of the right chambers was demonstrated, where the RV configured the apex of the heart, and presence of great hypertrophy of its walls and the moderator band, with moderately impaired systolic function. In the **table** the precise measurements are shown.

There was PA aneurysmal dilation (**Figure 2A**) in which a dissection flap forming the false lumen (**Figure 2B**) was observed. Both branches were dilated and a thrombus was detected in the right (**Figure 2B**). The pulmonary flow curve was type IV (**Figure 3A**) and the pulmonary acceleration time was severely decreased.

By subcostal view the existence of ostium secundum type ASD was demonstrated with right to left shunt shown by color Doppler. Saline contrast injection was further applied with large bubbles passing toward the left heart, thereby Eisenmeger's syndrome was demonstrated (**Figure 3B**).

The left cavities were rejected and displaced, the left ventricle was small with good global and segmental systolic function, and had an image in "D" in short-axis view, an expression of the increased RV diastolic pressure.

The angiography showed RV and RA dilation, with great dilation of PA and its branches, PA dissection with formation of a false lumen, and the true lumen (contrasted) was also clearly observed (**Figure 4**).

Medical treatment with oral furosemide (40 mg every 12 hours), sildenafil citrate (50 mg every 8 hours) and warfarin (2 mg per day) was administrated. Clinical improvement was obtained and the patient discharged with outpatient follow-up.

In an evolutionary echocardiogram at one year and a half of diagnosis, due to a new hospitalization, the PA diameter had increased to 90 mm.

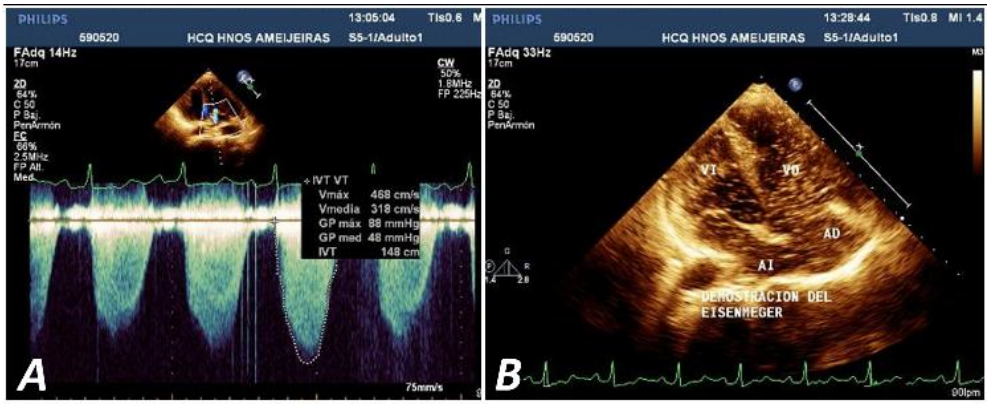

Figure 3. A. Continuous wave Doppler spectrum showing gradients of tricuspid regurgitation. Type IV pulmonary curve. **B.** Passing of saline contrast to left cavities after intravenous injection, demonstrating shunt inversion (Eisenmenger's syndrome). VI, left ventricle; AI, left atrium (by its acronym in spanish).

COMMENTS

Many of the patients with congenital heart disease, particularly those with significant left to right shunt, will develop pulmonary hypertension if they are not treated on time.

Persistent exposure to increased pressure determines changes in the pulmonary microvasculature

Table. Values of echocardiographic parameters measured at admission.

Echocardiographic Parameter	Measurement
BIDIMENSIONAL MODE	
Right atrium	
Longitudinal diameter	45 mm
Transverse diameter	32 mm
Area	14 cm²
Volume	44 ml
Right ventricle	
Basal diameter	43 mm
Mean diameter	36 mm
Longitudinal diameter	61 mm
Free wall	7 mm
Moderator band	8 mm
Main pulmonary artery	79,8 mm
Right branch	31,7 mm
Left branch	26 mm
ASD	
Ostium secundum type	20 mm
Inferior vena cava	
Expiratory diameter	24,2 mm
Diameter in inspiration	17,7 mm
Inspiratory collapse	< 50 %
Right ventricular function	
Systolic tricuspid annular plane excursion	14 mm
Lateral tricuspid annulus velocity by Doppler tissue	9,1 cm/s
Fractional shortening	15,7 %
Ejection fraction	34 %
DOPPLER HEMODYNAMIC	
Tricuspid regurgitation jet area	12,5 cm²
Maximum gradient	88 mmHg
Mean gradient	48 mmHg
Estimated pressure in RA	20 mmHg
Systolic pressure estimated in PA	108 mmHg
Mean pressure in PA	68 mmHg
Pulmonary acceleration time	46 ms

favoring increased pulmonary vascular resistance, in such a way that if the systemic resistance is overcome an inversion of the shunt will occur[5]. In response to the chronicity of this disorder, a degeneration of the tunica media of the pulmonary artery occurs that is associated with fragmentation of elastic fibers, which causes thinning and dilation of the vascular wall, which determines the increase in intravascular pressure. To this phenomenon, the wall stress is added, which favors intimal tear and causes the dissection of this artery[6,7], where, unlike the aortic dissection, the false lumen tends to produce a distal rupture creating a site of reentry[3,8].

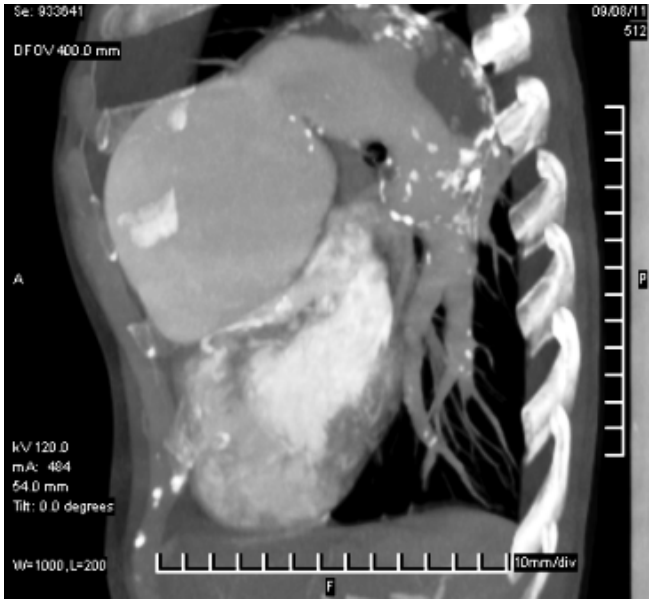

Figure 4. Tomographic angiography where the great dilation of the right cavities with huge expansion of PA and its branches, and the dissection of PA with formation of two lumens are observed. The true (contrasted) and the false.

Usually, in 80 % of cases, the dissection settles at PA without involving the branches, although it may rarely occur in isolation in one of the branches. It has been reported that prior aneurysmal dilation favors the appearance of dissection[3,9].

This disease is an extraordinary event that occurs equally in both sexes with an age range between 26 and 85 years and a peak incidence in the fourth to seventh decades of life. In young patients congenital heart diseases are the most common cause, while in the elderly there is a wide range of diseases of respiratory, inflammatory and autoimmune origin that can cause the dissection[10]. Among the most frequent clinical manifestations are chest pain and dyspnea at rest as cardinal symptoms[9].

In the last two centuries only 63 cases have been published, 8 of which have been diagnosed with life[11], which is due to technological advances in imaging modalities for diagnosis of pulmonary vascular disease, specifically advances in noninvasive methods such as echocardiography, computed tomography and magnetic resonance imaging. It should be noted that differences in application of each method depends on

availability, operator skills, individual anatomical variation, the location and distribution of intimal flap, as well as the inherent limitations of each imaging technique. Undoubtedly, echocardiography appears to be the first-line technique because of its easy accessibility and availability, low cost, accurate and immediate bedside diagnosis, and high positive predictive values [12-14].

It is appropriate to comment that the therapeutic approach in these patients is not well defined, some authors advocate conservative treatment with loop diuretics and phosphodiesterase type 5 inhibitors and more recently endothelin receptor inhibitors[15], and others advocate the use of reconstructive surgery and cardiopulmonary transplantation[14]. What is certain is the absence of medical practice due to the limited report of existing cases in the literature. However, we have the view that the decision should be taken by individualizing the patient to decide medical or surgical treatment depending on their status, comorbidity and the availability of invasive treatment, taking into account the risk-benefit ratio.

In this particular case, the conservative treatment was chosen and medical treatment was begun, considering that an indicated treatment option could be the cardiopulmonary transplantation, which is impossible in this patient due to her high comorbidity and all the conditions required to perform it. In order to improve her symptoms the previously mentioned pharmacological treatment was administered.

It is interesting to note that the patient has been kept alive since her diagnosis in August 2011 to the present. She was recently hospitalized with dyspnea at rest and an associated respiratory infection. In the evolutionary echocardiogram an increased diameter of the PA was found; an extremely rare fact that motivated the publication of this unusual and interesting case.

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