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Clinical case

Pulmonary artery dissection in Chagas heart disease

Disección de la arteria pulmonar en cardiopatía chagásica

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ABSTRACT. Objective: To describe the presentation of a pulmonary artery dissection to the Emergency Unit, the sequence of events, and a literature review. **Case report:** A 59-year-old female patient with known Chagas heart disease presented at the Emergency Unit with an event of sudden, stabbing chest pain and progressive dyspnea, evolving rapidly to cardiogenic shock. A chest radiograph reported severe widening of the mediastinum and cardiomegaly; therefore, a thoracic angiotomography was performed to rule out acute aortic syndrome, in which a dissecting pulmonary artery aneurysm was diagnosed. Transthoracic echocardiogram showed an intimal flap at the pulmonary artery and severe dilatation. The patient died in less than four hours after arrival at the emergency room without being able to access surgical treatment due to the rapid evolution. **Conclusion:** Pulmonary artery dissection is a rare disease, with a high rate of mortality. Optimal management requires a rapid diagnosis with multiple imaging techniques. There is a lack of information about this topic.
Keywords: Pulmonary artery dissection, Chagas heart disease, thoracic angiotomography.

RESUMEN. Objetivo: *Describir la presentación de una disección de arteria pulmonar a la Unidad de Urgencias, la secuencia de eventos y revisión de la literatura.* ***Reporte de caso:*** *Paciente femenino de 59 años con cardiopatía chagásica conocida, quien acudió a urgencias por un episodio de dolor torácico repentino punzante y disnea progresiva, que evolucionó rápidamente a choque cardiogénico. Una radiografía de tórax informó un ensanchamiento severo del mediastino y cardiomegalia; por tanto, se realizó una angiotomografía torácica para descartar síndrome aórtico agudo, en la que se diagnosticó un aneurisma disecante de arteria pulmonar. El ecocardiograma transtorácico mostró un colgajo de la íntima a nivel de la arteria pulmonar y una dilatación severa. La paciente falleció en menos de 4 horas desde su llegada a urgencias sin poder acceder a tratamiento quirúrgico debido a la rápida evolución.* ***Conclusión:*** *La disección de la arteria pulmonar es una enfermedad rara, con una alta tasa de mortalidad. El manejo óptimo requiere un diagnóstico rápido con múltiples técnicas de imagen. Falta información sobre este tema.*
Palabras clave: *Disección de la arteria pulmonar, cardiopatía chagásica, angiotomografía torácica.*

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INTRODUCTION

Pulmonary artery dissection (PAD) is a rare entity with high mortality. Since its first description by Helmbrecht in 1842,[1] less than 100 cases have been reported, most of them diagnosed post-mortem. Most of the patients die due to rapid-onset cardiogenic shock and sudden death. It usually occurs as a complication of chronic pulmonary hypertension.[2] In the largest case series reported, from 24 patients diagnosed alive, successful surgical repair was described only in seven subjects, attaining for the rapid evolution and fatal outcome.[2]

The present case report describes a middle-aged woman, previously diagnosed with chronic Chagas heart disease, with a dissecting

pulmonary artery aneurysm, cardiogenic shock and sudden death.

CASE PRESENTATION

The patient was a 59-year-old female, with a past medical history of arterial hypertension and Chagas heart disease.

She presented to the Emergency Unit with 1-month evolution of dysphonia, previously diagnosed as laryngitis, treated with prednisone 50 mg once a day for two weeks without improvement. One day prior to consultation, she suffers from a sudden onset of severe retrosternal pain, with stabbing characteristics, interscapular radiation, associated with progressive dyspnea at rest, that doesn't alleviate with analgesics.

On presentation, she was hemodynamically unstable, with pallor and acrocyanosis. Results on physical examination included a normal regular rhythm and a severe regurgitant tricuspid holosystolic murmur in addition to bilateral basal crackles. Arterial gasometry reports chronic respiratory acidosis and moderate acute respiratory distress syndrome (ARDS). Other laboratory tests are illustrated in *Table 1*.

Chest X-ray reveals marked mediastinal widening and vascular congestion with thickening at level of the aortic tract, an image suggestive of an aneurysm *(Figure 1)*.

As protocol to rule out acute aortic syndrome, a computed thoracic angiography was performed. In relation to the right ventricular outflow tract, an aneurysmal dilation of the pulmonary artery was reported, up to 5.3 cm diameter, with mass effect that compresses the aortic arch. It decreases the lumen to 1.3 cm causing pseudocoarctation. Also, an intimal flap was observed from pulmonary artery's origin, which extends to the right main pulmonary artery; true lumen gives rise to the left pulmonary artery, and incompletely to the right pulmonary artery, supplying only the right pulmonary upper lobe. Irrigation of the middle and lower right lobe origins from false lumen. A generalized cardiomegaly and a diffuse pulmonary mosaic pattern secondary to hypo perfusion were also described *(Figure 2)*.

Transthoracic, focused echocardiogram (TTE), describes a dilated cardiomyopathy, with reduced systolic function at rest, with a left ventricle ejection fraction of 25%, aneurysmal dilation of the pulmonary artery and the presence of an intimal flap. In addition, we identified a severe tricuspid regurgitation, with a peak velocity (Vmax) of 4.8 m/s, which stands to a high probability of pulmonary hypertension (PASP of 53 mmHg). There was no evidence of

Table 1: Laboratory tests at hospital admission.

Parameter	Value	Parameter	Value
White blood cells	17,000/mm^3	Glucose	96 mg/dL
Neutrophils	80%	Creatinine	0.77 mg/dL
Lymphocytes	13.7%	Uric acid	5.6 mg/dL
Eosinophils	2%	ALT	49 U/L
Monocytes	5.9%	AST	32 U/L
Hemoglobin	14.2 g/dL	CRP	24.9 mg/dL
Hematocrit	43.4%	ESR	62 mm/s
MCV	90.5 fL	LDH	290 U/L
MCH	29.6 pg	Sodium	139 mmol/L
MCHC	32.7 g/dL	Potassium	3.1 mmol/L
Platelets	234,000/μL	Phosphorus	3.7 mg/dL

MCV = median corpuscular volume; MCH = median corpuscular hemoglobin; MCHC = median corpuscular hemoglobin concentration; LDH = lactate dehydrogenase; CRP = C reactive protein; ALT = alanine aminotransferase; AST = aspartate aminotransferase; ESR = erythrocyte sedimentation rate.

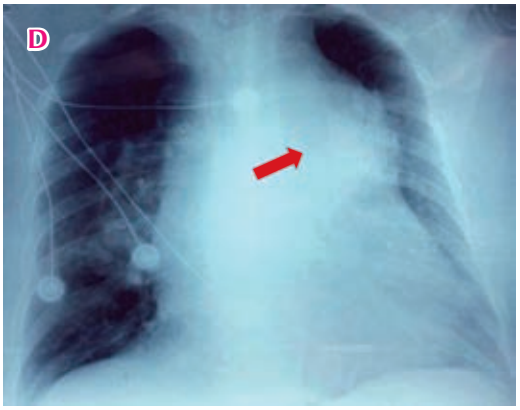

Figure 1: Chest X-ray. Cardiomegaly grade IV, mediastinal enlargement (red arrow) image suggestive of aneurysm.

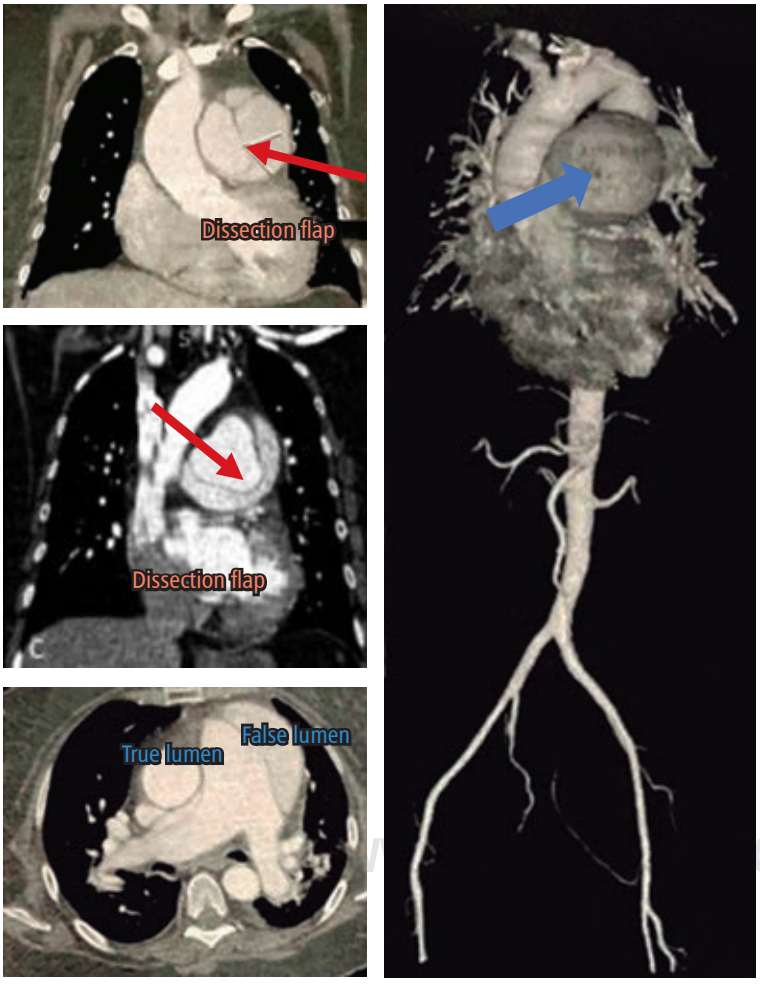

Figure 2: Thoracic angiography. Blue arrow indicates dissecting pulmonary artery aneurysm and its relationship with aortic arch (pseudocoarctation). Red arrow points to dissection flap.

pericardial effusion. Proximal aorta was normal *(Figure 3)*.

Due to its complexity, an evaluation by cardiovascular surgery was requested, recommending a surgical management; However, the patient presented a rapid evolution and dies in the Emergency Unit in less than 4 hours from arrival. For personal reasons, the autopsy was denied.

DISCUSSION

Pulmonary artery dissection (PAD) is an exceptionally rare condition with high mortality, frequently associated with pulmonary hypertension.[3,4] Occurs in both sex with a slight female predominance 1.2:1, with incidence peaks in the third and sixth decades of life.[5] The diagnosis is usually made in post-mortem studies due to the rapid progression that causes cardiogenic shock or sudden death.[6] The location of the dissection usually occurs in the major branches, predominantly in the trunk of the pulmonary artery in 72% of cases, followed by the intrapulmonary arteries in 10% and the involvement of the trunk and both main pulmonary arteries simultaneously in a minor percentage.[7]

Multiple causes of PAD are described, including chronic pulmonary hypertension leading to the development of a pulmonary artery aneurysm, congenital heart disease (mainly patent ductus arteriosus), chronic arteritis, pulmonary thromboembolism and rheumatic mitral valve stenosis.[8] Other less frequent causes are infections, trauma, atherosclerosis or connective tissue disorders.[9,10]

According to the literature, the clinical manifestations of PAD are nonspecific. The most frequent presentations are chest pain, dyspnea, cyanosis, hemoptysis, shock and death; dyspnea being the most common symptom in 82% of cases, while retrosternal chest pain and cyanosis have been described in 67% and 52% of cases, respectively.[11,12] Radiographic findings are nonspecific: cardiomegaly, dilatation of the trunk of the pulmonary artery and pleural effusion.[13] Computed tomographic angiography is a useful diagnostic tool in suspected pulmonary

artery dissection; however, its diagnostic performance has not been established due to the minority of reported cases.[14] Transthoracic echocardiography is an important imaging study in the initial approach by providing quantitative and qualitative information on cardiac function.

The optimal management of patients has not been defined due to the low number of cases in the literature. Based on anecdotal reports, surgical repair has been performed in few patients with variable results.[1] Medical treatment may include oxygen, analgesics, vasodilators, diuretics and inotropics.[15]

The patient has a medical history of chronic Chagas heart disease and hypertension who presented to the Emergency Unit with symptoms of atypical chest pain and physical examination with hemodynamic compromise; whose initial radiological findings led us to suspect an acute aortic syndrome; ruling out this cardiovascular emergency, the involvement of the pulmonary vasculature was documented in angiotomography, demonstrating a dissected aneurysm of the pulmonary artery of undetermined cause.

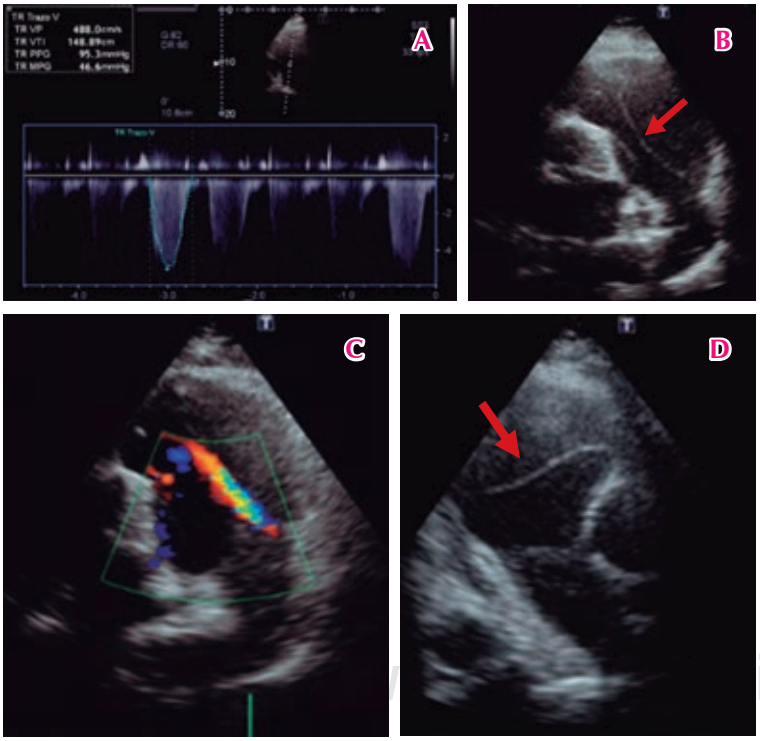

Figure 3: Transthoracic echocardiogram. **A)** Tricuspid regurgitation with a peak velocity (Vmax) of 4.8 m/s, and high echocardiographic probability of pulmonary hypertension. **B to D)** Great vessels short axis view. The arrow marks the dissecting flap in the pulmonary artery. **C)** Shows turbulence in Doppler color at the dissection point.

The series of events associated with aneurysmal dilation of the pulmonary artery is not known with certainty. Although there are no detailed studies in this regard, the development of aneurysms in the aorta, another large elastic artery, allows us to infer some common mechanisms. Thoracic aortic aneurysms are associated with classic cardiovascular risk factors, chest trauma, vasculitis (and other autoimmune diseases) and genetic factors in up to 25% of cases. Multiple genetic pathways are related to the development of aortic aneurysms and pulmonary hypertension, either in high penetrance familial forms (Marfan, Loeys Dietz, Ehlers-Danlos syndromes) or in sporadic mutations. The latter, perhaps more frequent in general population, requires a higher influence of environmental factors for phenotypic expression, among them, chronic left ventricule afterload alterations.[16]

In the case of our patient, hemodynamic overload due to type II pulmonary hypertension could favor the phenotypic expression of the pulmonary artery aneurysm, with unknown pathogenic mutations. Finally, case reports associate the chronic use of steroids with the presentation of aortic and pulmonary dissection, contributing to degeneration of the vascular media. Although the duration of therapy was short (only two weeks), high doses of corticosteroids (0.5 mg per kilogram of body weight) may be a predisposing factor in the presentation of this patient.[16]

Despite the fact that successfully surgical interventions are reported, the high risk that the intervention entails impoverishes the prognosis of patients, showing that even in conservative management despite optimal medical treatment, mortality is about 100%.[2]

We consider this case report of high academic value, given the unusual symptoms at presentation (dysphonia), treated erroneously as an upper respiratory infection. In addition, the high similarity with an acute aortic syndrome, with whom it shares a high mortality despite an adequate treatment instituted.

CONCLUSIONS

Pulmonary artery dissection is an extremely rare disease, generally with fatal outcome. Due to the few reported cases in the literature, there is a lack of management and treatment guidelines for this entity with high mortality, so the approach is based on case reports and clinical experiences. More studies are needed on this entity to improve the diagnostic and therapeutic approach.

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