

Case 1 - A 33 Year-Old Man with Effort Dyspnea and Syncope who Presented Sudden Worsening of the Dyspnea

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The patient started to present shortness of breath, which initially appeared at great efforts and progressed until it was also triggered by mild efforts. Fatigue and asthenia were associated to this picture. Four months before, the individual had presented several episodes of syncope triggered by physical exertion.

The patient sought medical assistance at other services. The medical evaluation included laboratory assessment. The Doppler echocardiogram performed at that time showed dilatation of the right cardiac chambers and the pulmonary artery, marked tricuspid insufficiency and pulmonary hypertension. The systolic pressure of the pulmonary artery was estimated at 57 mmHg.

The patient was also submitted to a coronary angiography, which did not show luminal obstructions in the coronary arteries. The patient was prescribed 0.25 mg of digoxin and 40 mg of furosemide every three days.

A few days before the patient was admitted at this hospital, he presented worsening of the dyspnea, which started to occur even at rest and he sought medical assistance (November 26, 2006). At physical examination (November 26, 2006), he presented tachypnea, with cold extremities and signs of poor peripheral tissue perfusion. The heart rate (HR) was 95 bpm, blood pressure (BP) increased to 100/60 mmHg and the jugular venous pressure was increased. The pulmonary auscultation was normal; the second heart sound was accentuated in the pulmonary area and there was a 4/6+ intensity systolic murmur in the tricuspid area. The abdomen was little dilated and there was painful hepatomegaly, as well as mild lower-limb edema.

The electrocardiogram (ECG) performed on November 26, 2006, showed sinus rhythm, HR of 75 bpm, SÂQRS +120° tending forward, false left atrium overload (negative final slow P wave inscription at V1) and right ventricular overload, compatible with systemic pressures in this chamber (Figure

1). The chest x-ray (November 26, 2006) disclosed signs of pulmonary trunk dilatation.

The echocardiogram (November 27, 2006) disclosed a left atrium diameter of 37 mm, aorta diameter of 30 mm, septal thickness of 8 mm and posterior wall thickness of 8 mm; left ventricular diameter of 32 mm/20 mm (diastolic/systolic) and a left ventricular ejection fraction of 69%. The right ventricle was dilated and presented marked hypokinesis. There was marked tricuspid insufficiency and the pulmonary systolic pressure was estimated at 90 mmHg.

The laboratory assessment (November 26, 2006) showed: hemoglobin 8.9g/dl, hematocrit 31%, MCV 65 fl; leukocytes 10500/mm³ (neutrophils 66%, eosinophils 1%, lymphocytes 17% and monocytes 16%), platelets 208000/mm³, creatinine 1.63 mg/dl, urea 53mg/dl, glycemia 97 mg/dl, total bilirubin 2.8 mg/dl and direct bilirubin 1.79 mg/dl, alkaline phosphatase 195 UI/l, gamma GT 217 UI/l, aspartate aminotransferase (GOT) 60 UI/l, alanine aminotransferase (GPT) 100 UI/l, lactate dehydrogenase 376 UI/l, D-dimer 2704 ng/ml, brain natriuretic peptide 986 pg/ml, ionized calcium 1.08 mEq/l, chlorides 104 mEq/l, magnesium 1,9 mEq/l, lactate 151 mg/dl or 17 mmol/l (normal <20 mg/dl) or < 5 mmol/l), potassium 4.9 mEq/l, sodium 140 mEq/l, bicarbonate 6.3 mEq/l, and anion gap 38.6 mEq/l. Gasometry, with the patient using an O₂ catheter (November 27, 2006; 3h34) disclosed pH 7.27, pCO₂ 14.3 mmHg, O₂Sat 96.8, bicarbonate 6.3 mEq/l, excess of base (-) 19.4 mEq/l.

The diagnoses of pulmonary hypertension of unclarified etiology and recent pulmonary thromboembolic episode were made. The patient received a solution of 0.9% sodium chloride, 40 ml-25% glucose solution (due to an initial capillary glycemia of 40 mg/dl, which persisted at this level and therefore, another 40 ml of the glucose solution at 25% was administered) and 8.4% sodium bicarbonate. A new gasometry, with the patient wearing an O₂ mask (November 27, 2006) showed a pH of 7.11, pCO₂ 12.3 mmHg, pO₂ 123 mmHg, Sat O₂ 96.1%; bicarbonate 3.7 mEq/l and excess of base -24.7 mEq/l. The intravenous administration of streptokinase was initiated. The patient presented cardiorespiratory arrest with pulseless electrical activity, which was unresponsive to resuscitation maneuvers and died (November 27, 2006).

Key Words

Pulmonary hypertension; heart failure; pulmonary embolism; cardiogenic shock.

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Clinical aspects

This was a young male patient, with a picture of worsening of the dyspnea two days before hospital admission. He had been complaining of progressive dyspnea on physical exertion, associated to fatigue and asthenia for a year; four months before, he had presented episodes of syncope on physical exertion.

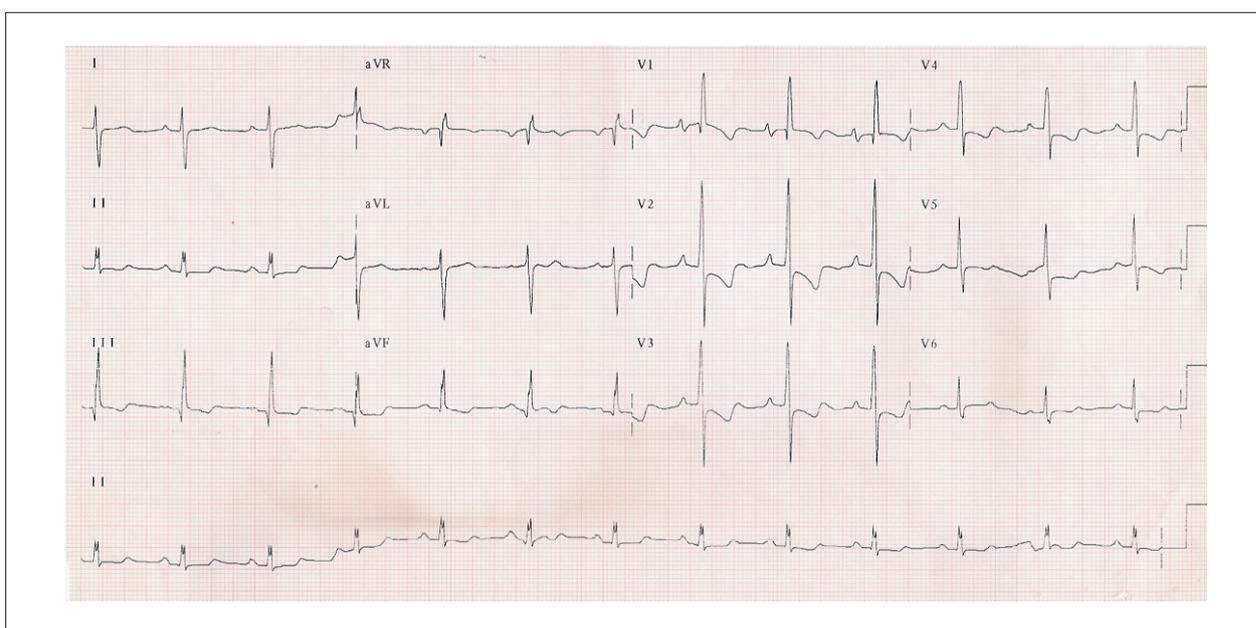


Figure 1 - ECG. False left atrium overload. It is a right atrium and right ventricle overload compatible with systemic pressures in this chamber.

At the time, digoxin and furosemide were initiated, due to the evidence of pulmonary hypertension; however, there was no investigation regarding the etiology. The patient developed a picture of pulmonary hypertension, progressive dyspnea, lower-limb edema and liver congestion.

The patient was admitted at the hospital with a picture of low output and signs of right heart failure and developed cardiorespiratory arrest with pulseless electrical activity, which was unresponsive to resuscitation maneuvers and died one day after hospital admission.

At least two points must be considered in this clinical case. The first one is about the assessment of the underlying clinical picture, emphasizing the diagnosis and the initial investigations.

Patients with suspected pulmonary hypertension, as in the present case, with a suggestive clinical and echocardiographic picture, must be submitted to extensive diagnostic assessment, in order to confirm the suspected pulmonary hypertension and identify its underlying cause. The diagnosis attained through a single examination followed by treatment prescription is an attitude that cannot be justified in any circumstance at present. The patients must be followed or at least advised by professionals with experience in the treatment of the disease.

Pulmonary hypertension is defined as a hemodynamic consequence of progressive vascular alterations of varied etiologies that occur in the pulmonary territory. Quantitatively, it is defined as a pulmonary artery pressure > 25 mmHg at rest or > 30 mmHg on exertion, measured by direct heart catheterization.¹

According to the World Health Organization, the pulmonary hypertension must be divided in five groups, based on their etiology.² Patients from the first group have

a tendency to present pulmonary artery hypertension. This group includes patients with sporadic idiopathic pulmonary hypertension, familial idiopathic pulmonary hypertension and those that present diseases located in the arteriolar territory, such as collagen disease, congenital cardiac disease and portal hypertension associated to HIV and anorexigenic drugs.

The other groups are classified as: pulmonary venous hypertension, caused by diseases that affect the left atrium and/or ventricle or left valvulopathy; pulmonary hypertension associated to hypoxemia and respiratory system disease, such as interstitial disease, chronic obstructive disease and sleep apnea; pulmonary hypertension caused by chronic thrombosis or embolic disease, affecting both the proximal and distal vasculature; and pulmonary hypertension caused by inflammation, mechanical or extrinsic obstruction of the pulmonary vasculature, such as sarcoidosis and histiocytosis.

A detailed anamnesis, thorough physical examination and careful interpretation of the ECG and chest x-ray are the initial steps for the clinical diagnosis of pulmonary hypertension, in addition to the sequence of examinations to be considered. In this assessment, it is crucial to rule out some disease that can be surgically treated, such as cardiac defects (shunts, obstructions and anomalous pulmonary venous drainage) and chronic pulmonary thromboembolism, or those that can receive specific treatment, such as schistosomiasis, collagenosis and pulmonary vasculitis.

Although it is difficult at the Emergency Room, to obtain the clinical history of the patient, detailed from childhood, is fundamental and must contemplate all etiological possibilities. The pulmonary hypertension can occur "silently" due to cardiac and systemic diseases or diseases in the lung parenchyma and vasculature. Its diagnosis is usually made at advanced phases of the associated diseases, responsible for

most of the symptoms. As with our patient, the most common symptom is intolerance to physical exertion (effort dyspnea) and fatigue due to low cardiac output, with a progressive characteristic and indicating secondary right ventricular dysfunction. Syncope or pre-syncope are worrying symptoms, as they indicate low cardiac output due to right ventricular dysfunction and/or ventricular arrhythmia. Although not present in this case, some symptoms can indicate the etiology of the pulmonary hypertension, such as orthopnea, present more often in patients with post-capillary or mixed pulmonary hypertension and also verified in patients with chronic obstructive pulmonary disease; nocturnal paroxysmic dyspnea, highly suggestive of venocapillary pulmonary hypertension; precordial pain of the angina type triggered by exertion, which can indicate an ischemic cardiopathy or a right ventricular subendocardial ischemia, due to the decreased coronary perfusion flow and pressure; hemoptysis, which is an unusual symptom and can alert to the possibility of pulmonary thromboembolism and pulmonary infarction or advanced mitral stenosis, etc.

The physical examination is extremely useful for the diagnosis of pulmonary hypertension and its possible cause. As in the patient in question, the increase in the intensity of the pulmonary component of the second heart sound is the most consistent sign, regardless of its etiology and its transmission to the heart apex also indicates pulmonary hypertension. The presence of jugular stasis and the tricuspid and pulmonary insufficiency murmur are findings that are secondary to pulmonary hypertension. Although not present in this patient, some signs are noteworthy or indicate an etiologic diagnosis: murmurs can also indicate mitral stenosis, mitral insufficiency and obstruction of the left ventricular outflow tract (aortic, sub-aortic or supra-aortic stenosis), causing post-capillary pulmonary hypertension; auscultation of wheezing, rhonchi and crackling rales suggest pulmonary diseases (bronchitis, emphysema, asthma, pulmonary fibrosis); the occurrence of Raynaud's phenomenon, of skin lesions and arthritis indicate collagenoses; and signs of chronic hepatopathy might suggest hepatopulmonary syndrome or schistosomiasis etiology.

All patients with suspected pulmonary hypertension, after a detailed anamnesis and clinical assessment, must be submitted to a series of complementary examinations. The objective is to confirm the pulmonary hypertension, evaluating its severity, clinical implications, hemodynamic characterization and mainly, trying to establish the etiology and allow the therapeutic planning.

Additional examinations are necessary to elucidate the etiology or coexisting diseases in selected individuals, as in the patient in question: electrophoresis of hemoglobin to rule out sickle-cell disease; antinuclear antibodies and rheumatoid factor to rule out collagenoses; coagulation tests such as lupus anticoagulant factor; anti-thrombin III, C and S proteins to evaluate the hypercoagulability state and serological tests for HIV as well as B and C hepatitis viruses. If we knew the patient's origin and schistosomiasis was suspected, the parasitological and serological evaluations as well as rectal valve biopsy and presence of portal hypertension would be important.

The ECG presented a characteristic pattern with signs of right chamber overload, right QRS axis deviation ($>90^\circ$), right

bundle-branch block, wide R waves at V1, V2 and deep ones at V5, V6, with T-wave and ST-segment alterations. Although it does not apply to the current case, the careful analysis of the ECG can suggest some etiologies for pulmonary hypertension, such as mitral stenosis, congenital cardiopathies and myocardiopathies.

The chest x-ray supported the diagnosis of pulmonary hypertension, demonstrating enlargement of the pulmonary artery and the right cavities. There can also be attenuation or disappearance of the pulmonary veins in the peripheral site. The radiography is usually very useful to rule out or suggest pulmonary causes, such as chronic obstructive pulmonary disease, pulmonary fibrosis, interstitial disease and granulomatous diseases.

The echocardiogram, in this case, was very useful for the diagnosis of pulmonary hypertension, as it assessed the cardiac structure and function, quantifying the tricuspid reflux as accentuated. It also ruled out the presence of intracardiac communications or in the vessels of the base and post-capillary causes of pulmonary hypertension, such as left atrial myxoma, mitral stenosis and other cardiac diseases.

In spite of the diagnostic limitations found in this case, we have a case of a patient with a picture of pulmonary hypertension of undefined etiology that presented a fast and dramatic evolution, compatible with a picture of idiopathic arterial pulmonary hypertension. That is a rare entity, of which prevalence is estimated at 1-2 cases per million individuals in the general population. It usually affects young individuals between the third and fourth decades of life, as in the present case, although there is a predominance of the female sex. Although the diagnosis of idiopathic arterial pulmonary hypertension is an exclusion diagnosis, it was possible to attain it with high probability. Thanks to imaging methods, such as transesophageal echocardiogram, ventilation-perfusion scintigraphy, high-resolution helicoidal tomography, angiography by magnetic nuclear resonance and functional tests, we are capable of excluding almost all diseases that cause pulmonary hypertension.

The second point to be discussed is the assessment and treatment of the possible causes of decompensation of the underlying disease.

The picture at the ER admission consists of low output syndrome without left ventricular dysfunction, characterized by signs of poor peripheral perfusion, clear pulmonary auscultation and absence of a third heart sound at the physical examination; laboratory assessment showing a lactate level of 151 with severe metabolic acidosis and increased anion gap; pulmonary hypertension syndrome with right ventricular failure; increased BNP level; laboratory assessment showing indirect signs of hepatic congestion and echocardiogram disclosing right ventricular failure without left ventricular abnormalities.

Considering this dramatic picture, the differential diagnosis of causes of obstructive shock is necessary, with the most common being hypertensive pneumothorax, cardiac tamponade and diseases that increase the pressure on the pulmonary arterial bed. For this patient, the diseases that increase the pulmonary artery pressure are the most probable diagnoses, as the physical examination with pulmonary auscultation and chest x-ray, with no alterations suggestive of pneumothorax and the

echocardiogram without signs of pericardial effusion practically rule out the two other diagnostic possibilities.

Among the causes of exacerbated chronic pulmonary hypertension, massive acute pulmonary thromboembolism, acute pulmonary arterial thrombosis and the natural evolution of the underlying disease are the main hypothesis for patients with no other symptoms, disclosed by the clinical assessment and complementary examinations, which could define a specific condition of the systemic disease and secondary pulmonary involvement (such as congenital, rheumatic, infectious diseases and other causes of pulmonary hypertension reported above).

The massive pulmonary thromboembolism, characterized by right ventricular dysfunction and shock and/or hypotension, as well as the acute pulmonary thrombosis, presents high mortality and its level of suspicion must also be high, so that the treatment can be instituted early. The clinical history of chronic dyspnea, as a consequence of a picture of pulmonary hypertension of undefined etiology and acute exacerbation in the last couple of days – as in the present case – strongly suggests acute pulmonary thromboembolism. The recent guidelines recommend different approaches for its diagnosis, varying according to the availability of the methods used at each service and the clinical condition of the individual.³

Beside examinations such as complete biochemical assessment, complete blood count, myocardial necrosis markers, BNP, D-dimer by ELISA method (Enzyme-Linked ImmunoSorbent Assay), arterial gasometry, ECG, chest x-ray, echocardiogram and lower-limb Doppler ultrasound are very useful, although of little specificity for the diagnostic rationale and decision making. Such procedures show important findings in patients hemodynamically unstable and incapable of undergoing examinations for a final diagnosis of massive pulmonary thromboembolism, such as V/Q pulmonary scintigraphy, helicoidal computed tomography (CT) and pulmonary arteriography. Among these assessments, when possible, the helicoidal CT is the most indicated one, due to the fact that it is relatively easy to perform when compared to the other means, in addition to the sensitivity and specificity > 95%. As we know, the mortality due to massive pulmonary thromboembolism usually occurs within the first hours of picture evolution. As soon as its diagnosis is suspected or confirmed by the aforementioned information, the therapeutic, with general support measures, volemic expansion and inotropic drugs must be started together with other measures that promote the lysis of the thrombus. The standard treatment consists of thrombolytic agents. The most often used ones are streptokinase and the recombinant tissue plasminogen activator (r-tPA). The surgical embolectomy is indicated in shock refractory to clinical treatment and has a very high mortality rate.

The natural evolution of the disease is also a probable cause, considering that the patient already presented, at the time of the diagnosis, a picture of advanced disease. Poor prognosis criteria, such as NYHA functional class IV, characterized by dyspnea on mild effort, syncope triggered by exertion and echocardiographic criteria with right ventricular and pulmonary artery dilatation and accentuated tricuspid insufficiency, confer to this patient a mean survival of six months, compatible with the evolution of the disease in the reported case. Other criteria of poor prognosis present

in the acute as well as in the chronic phase of the disease are increased levels of BNP and D-dimer, as also seen in this case.

We know that idiopathic arterial pulmonary hypertension is an incurable disease, of which mean time of survival after the diagnosis is approximately two years and a half, according to the National Institutes of Health (NIH). The use of oral anticoagulants increases survival by three years and the patients responsive to calcium-channel blockers have a significantly longer survival, when compared to the nonresponsive ones. The chronic use of epoprostenol in patients with functional class III or IV is associated with a significant decrease in mortality and the need for transplant, with a 5-year survival of 54%, twice that of patients from the control group (27%). The predictors of worse prognosis are the intensity of pulmonary hypertension, the degree of right ventricular dysfunction, the low cardiac output, the degree of peripheral desaturation and the low tolerance to physical exertion. Most of the patients die due to right ventricular failure (cardiogenic shock): of this total, approximately 10% have a sudden death.³

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Diagnostic Hypothesis

Cor pulmonale due to pulmonary hypertension caused by massive pulmonary thromboembolism.

Necropsy

The necropsy showed cardiac hypertrophy and accentuated dilatation of the right chambers, mainly in the right ventricle, as well as thrombosis in the right auricle (Figure 2). There were no lesions suggestive of endomyocardial fibrosis in either ventricle. The pulmonary trunk and the central pulmonary arteries presented dilatation and irregularities on the intimal surface, consistent with atherosclerosis, suggesting accentuated hypertension in the pulmonary circulation (Fig. 3).

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The macroscopic assessment showed signs of congestive heart failure, represented by chronic passive congestion, with “nutmeg liver”, in addition to areas suggestive of hepatic cirrhosis and mild splenomegaly. The histological analysis showed granulomas involving parasite eggs with characteristics of *Schistosoma mansoni* present in the lungs and liver. In the latter, in addition to central-lobular sinusoidal ectasia due to chronic passive congestion, there was also stellate-pattern portal fibrosis and it sometimes formed porto-portal and porto-central bridges, consistent with the sequelae of hepatic schistosomiasis (Fig. 4).

In the lungs, the granulomas were scarce and were associated to frequent obstructive arterial lesions, consistent with severe pulmonary hypertension, grade III (total occlusion of the arteriole lumen) and IV lesions (plexiform-type lesions) of Heath-Edwards; acute pulmonary thromboembolism was also verified (Fig. 5). This was detected in the upper and lower lobes of the left lung, with probable origin in the right auricle, mentioned above, and considered as the cause of the decompensation of the

Anatomopathological Session

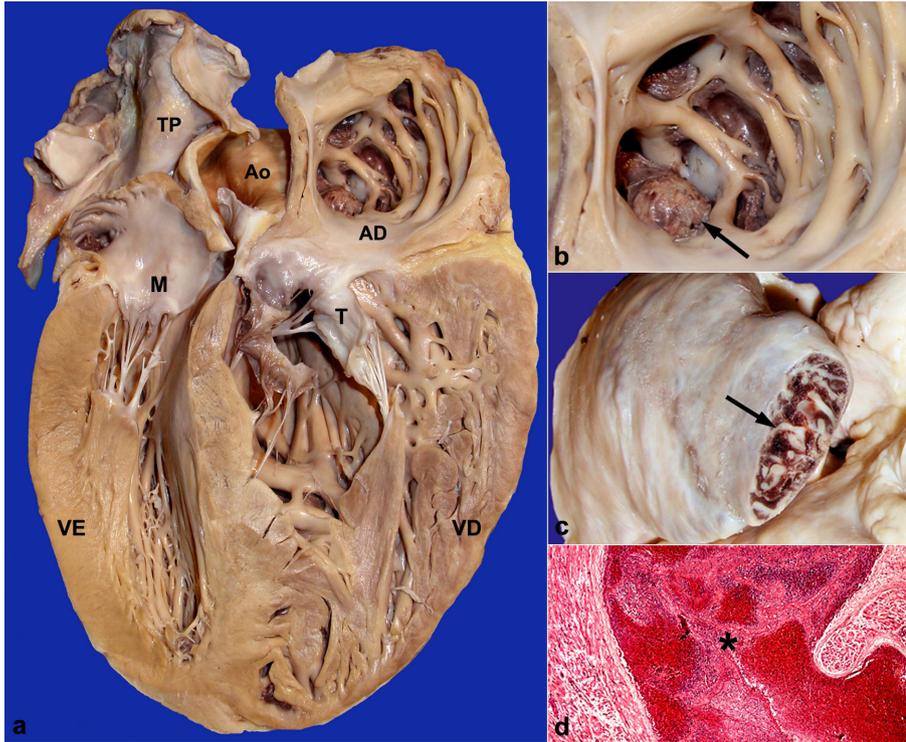


Figure 2 - Heart images. In (a) we observe a large volume of the right chambers, mainly in the right ventricle (RV), with marked hypertrophy of the wall and cavitory dilatation in relation to the left ventricle (LV). The right atrium (RA) is better observed in Figure (b), which shows the presence of thrombosis (arrow), permeating the pectineal musculature and filling up the right auricle (arrow), of which tip was cut (c). Histological section of the auricle (d) showing fibrinohematic thrombus (*), filling up the auricular cavity (Hematoxylin & eosin; 5X magnification). PT - pulmonary trunk, Ao - aorta, M - mitral, T - tricuspid.

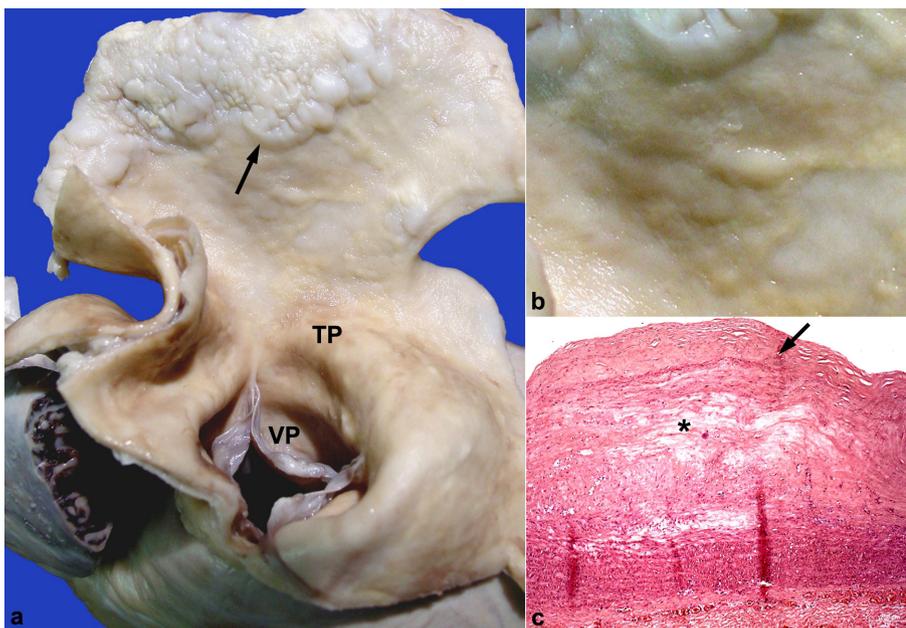


Figure 3 - Images of the pulmonary trunk and part of the open central pulmonary arteries. In (a), we observe the trunk dilatation and the intense irregularity on the intimal surface, with fibrotic plaques (arrow) and delicate yellowish nodularity – better observed in (b) – suggestive of atherosclerosis that occur in pulmonary hypertension. The histological analysis (c) confirms the atherosclerosis, where the clear spaces are the negative image of fat accumulation (*) covered by a fibrous layer (arrow). (Hematoxylin & eosin; 5X magnification).

chronic *cor pulmonale*, i.e., of the right ventricular dilatation.

The cause of death was cardiogenic shock, with consequent shock lung and cerebral edema, with cerebral amygdala grooving.

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Anatomopathological diagnosis

Compensated hepatosplenic schistosomiasis; hypertensive vascular-pulmonary, with marked vascular-pulmonary alterations, indicating severe pulmonary hypertension;

chronic *cor pulmonale*, decompensated by recent pulmonary thromboembolism; congestive cardiac failure, predominant on the right side; cardiogenic shock.

Comments

The World Health Organization (WHO) estimates that schistosomiasis affects around 200 million people in 76 countries.⁴ In Brazil, schistosomiasis, which is also known as bilharzia, bilharziosis or snail fever, seems to infect around 6 million people that inhabit mainly the northeastern states and

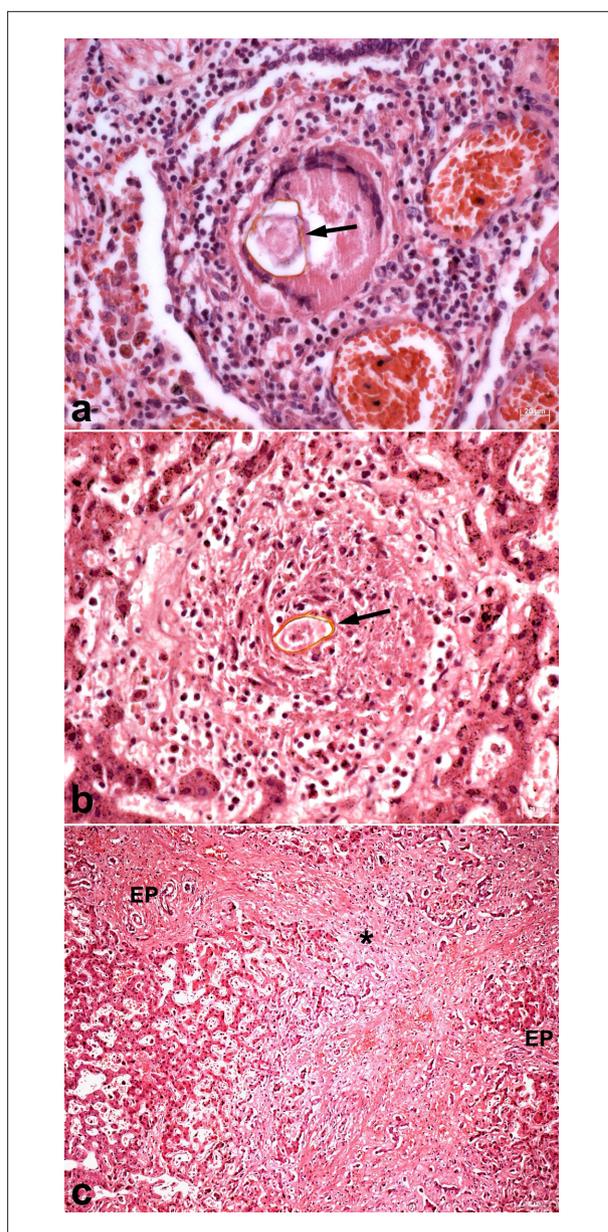


Figure 4 - The histological analysis showed granulomas involving parasite eggs, with characteristics of *Schistosoma mansoni* (arrows) in the lung (a) and liver (b). In the liver (c), the portal spaces (PS) presented expansion by fibrosis, forming, in certain areas, porto-portal bridges (*) (Hematoxylin & eosin; magnification of 40x, 40x and 5X, respectively).

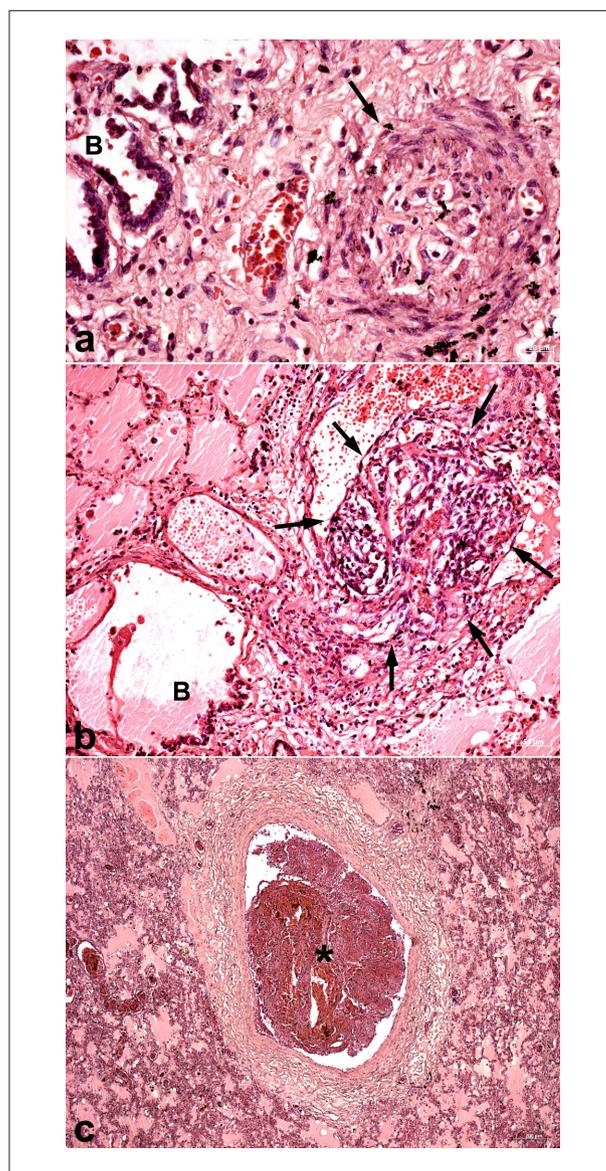


Figure 5 - The pulmonary histological analysis showed severe obstructive arteriole lesions, with multiple lesions as in (a), with total occlusion of the arteriole lumen (arrow) of the respiratory terminal bronchiole (B) and as in (b), with plexiform lesions (between the arrows), which resemble a renal glomerule, in respiratory bronchiole arteriole (B). The image (c) shows the recent pulmonary thromboembolism (*) in small-caliber pulmonary artery, present in the case (Hematoxylin & eosin; magnification of 40x, 20x and 5x, respectively).

the state of Minas Gerais.⁵ This is an important socioeconomic indicator related to poverty. Data from Ministry of Health regarding the year 2005 still indicate several areas of high, medium and low endemicity of schistosomiasis in Brazil, as well as multiple foci of the disease present in several other states located along the Brazilian Atlantic shore, including Sao Paulo.⁶

Recalling the life cycle of the *Schistosoma mansoni*, it involves two hosts. Its eggs are eliminated in human feces and upon reaching the water, the eggs hatch, originating parasites that enter and infect the intermediate host: the snail of the *Biomphalaria* genus. Inside the snail, the parasite develops, originating cercariae; in the water, they penetrate the skin of persons who are wading, swimming, bathing, or washing in contaminated water. After the penetration, the cercariae are called schistosomules. They enter the venous circulation; reach the lungs, heart, mesenteric arteries and portal system, where the sexual maturation occurs, originating male and female worms. They descend to the mesenteric veins, to the thinnest branches, where they reproduce, and the eggs, after passing through the submucosa into the intestinal lumen, are eliminated in the feces, thus closing the cycle.^{7,8}

The pathogeny of the disease is complex and involve the parasites and their eggs; the immunological phenomena, dependent on the host and the vessel alterations,^{7,8} which represent the parasite habitat. The anatomoclinical forms of the disease are divided in acute and chronic phases. In the acute phase, the penetration of the skin by the cercariae can be asymptomatic or result in cercarian dermatitis – a sensitivity reaction in individuals that have been infected before, with a rash-like manifestation, papular eruption, erythema, edema and pruritus. The dissemination of the larvae in the blood and their division in the lungs and later, in the liver, activates the immune system, causing fever, malaise, headaches, asthenia, abdominal pain, bloody stools, dyspnea, hemoptysis, arthralgia, lymph node enlargement and splenomegaly, in addition to a set of symptoms known as Katayama syndrome (also called acute schistosomiasis or Katayama fever).^{6,7}

The chronic phase, or chronic schistosomiasis, starts six months after the infestation, and can last for years. The parasite has a long survival, usually varying from 5 to 15 years; a case of a 25-year survival was verified in a patient that had left the infected area. In this phase, the signs of disease progression can appear in several organs and can reach extreme degrees of severity, such as pulmonary and portal hypertension, ascites and esophageal varicose vein rupture. The clinical manifestations vary, depending on the location and intensity of the parasitic infestation, the individual's capacity of response or the instituted treatment. The anatomoclinical forms of the chronic phase are: asymptomatic, intestinal (Type

I), hepatointestinal (Type 2), compensated hepatosplenic (Type III) and decompensated or complicated (Type IV - which includes the most severe forms), vasculopulmonary (hypertensive, cyanotic), tumoral (or pseudoneoplastic) and ectopic forms. However, eggs and adult worms of the parasite can be found in any organ or tissue of the human body, such as the brain, testicles and ovaries, among others. Among the ectopic forms, the most severe is the neuroschistosomiasis (schistosomatic myeloradiculitis).^{5,6}

Regarding the anatomopathological alterations, in acute schistosomiasis there is a miliary dissemination of eggs and necrotic-exsudative granulomas are observed, mainly in the liver and intestines (diffuse catarrhal enterocolitis) and splenic lymphoid hyperplasia. In the chronic phase, there is chronic catarrhal enterocolitis in the intestine (edema, granulation, petechia, mucus and ulcerations). The reaction is diffuse, with lymphocytary infiltrate and eosinophilia; sometimes, it promotes a hyperplastic or pseudotumor reaction, with polyp-like formations. In the liver, the basic lesion is the vascular one: inflammatory and fibrotic reaction around the portal tree. In the mild forms, there are sparse periovular granulomas, variable lymph-plasmocytary portal infiltrate, moderate portal fibrosis, formation of thin septa and schistosomatic pigment.

More accentuated hepatic forms produce the so-called Symmers' fibrosis (clay pipe-stem cirrhosis on the cross-section and nodular external surface). The vascular lesions are mainly caused by the eggs, with intra- and extra-arterial granulomas.^{6,7} The hepatic and the intestinal involvement are the most frequent and important ones, followed by the pulmonary involvement, of which the vascular lesion and consequent pulmonary hypertension lead to the development of *cor pulmonale*, as in the case described here. The lungs are affected in around 15.5% of the patients.⁷ The prevalence of pulmonary hypertension associated to schistosomiasis is variable. However, it is estimated that it can occur in up to 30% of patients in the hepatosplenic form.⁸ Incidentally, in a study with 123 patients that presented a diagnosis of pulmonary hypertension in two reference centers in the city of Sao Paulo, 30% had the disease associated to schistosomiasis.⁹

Thus, schistosomiasis in our country is still a disease to be considered among the differential diagnoses during the routine care of patients and the compulsory notification of cases in non-endemic areas is mandatory, as established in the Decree MS/GM number 2.325, of 12/08/03, as well as of all severe forms in the endemic areas and all cases in the endemic areas with isolated foci (states of Para, Piaui, Rio de Janeiro, Sao Paulo, Parana, Santa Catarina, Goias, Distrito Federal and Rio Grande do Sul).⁶

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