

Biventricular heart failure secondary to calcified tuberculous constrictive pericarditis: a case report and review

Falla cardíaca biventricular secundaria a pericarditis constrictiva calcificada de origen tuberculoso: reporte de un caso y revisión

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Key words:

Pulmonary tuberculosis, heart failure, calcified pericardium.

Palabras clave:

Tuberculosis pulmonar, insuficiencia cardíaca, pericardio calcificado.

ABSTRACT

A 41-year-old man with a medical history of pulmonary tuberculosis presented to the emergency department complaining of progressive dyspnea, lower limbs edema and increased abdominal volume of three months of evolution. He was diagnosed of heart failure by clinical findings. However imaging tests showed a heavily thickened calcified pericardium and bilateral pleural effusion. A skin tuberculin test was performed, giving a positive result. The diagnosis of biventricular heart failure secondary to calcified tuberculous pericarditis was reached. The patient rejected surgical intervention and conservative treatment for heart failure as per guidelines was implemented.

RESUMEN

Un hombre de 41 años con antecedente de tuberculosis pulmonar se presentó en el servicio de urgencias refiriendo un cuadro de disnea progresiva, edema de miembros inferiores e incremento del volumen abdominal de tres meses de evolución. El paciente fue diagnosticado de insuficiencia cardíaca por los hallazgos clínicos. Sin embargo estudios de imagen mostraron un pericardio importantemente calcificado y derrame pleural bilateral. Se realizó la prueba de tuberculina, siendo positiva. Se llegó al diagnóstico de insuficiencia cardíaca biventricular secundaria a pericarditis constrictiva de origen tuberculoso. El paciente rechazó la intervención quirúrgica y se implementó tratamiento conservador para falla cardíaca sustentado en guías.

INTRODUCTION

Constrictive pericarditis is the result of a chronic inflammatory process that leads to scarring, thickening, fibrosis and calcification of the pericardium which leads to impaired diastolic filling of the heart. The most common causes are mediastinal radiation, chronic idiopathic pericarditis, and after cardiac surgery in developed countries. However, tuberculosis is still the most frequent cause in the developing and underdeveloped world, as well as, in immunocompromised patients.^{1,2} It can occur after virtually any pericardial disease process but the risk of progression is related to the etiology; being low (1%) in viral and idiopathic pericarditis, intermediate (2-5%) in immune-mediated

pericarditis and neoplastic pericardial diseases and high (20-30%) in bacterial pericarditis, especially purulent pericarditis.³

In constrictive pericarditis the diastolic filling of the ventricles is impaired and clinical manifestations are generally those of right heart failure with preserved left ventricular function, at least in the first stages of the disease. Symptoms include fatigue, peripheral edema, breathlessness, passive hepatic congestion, nonspecific abdominal complaints; and as the disease progresses, abdominal swelling secondary to ascites, jaundice, generalized edema and cardiac cirrhosis. Other important signs are the recurrent pleural effusions and syncope, due to low output status secondary to reduced preload.³⁻⁷

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The jugular veins are distended. The normal inspiratory drop in jugular venous distention may be replaced by a rise in venous pressure (Kussmaul's sign) due to a right ventricle unable to accommodate the blood proceeding from the venous return, being into a fixed space, delimited by the stiff pericardium. Kussmaul's sign is also present in cases of severe right heart failure, especially in association with tricuspid regurgitation. Pulsus paradoxus is present when systolic pressure decreases more than 10 mmHg during inspiration, and it is secondary to the shift of the interventricular septum to the left, impeding an adequate filling of the left ventricle. The classic auscultatory finding of pericardial constriction is a pericardial knock that occurs as a high-pitched sound early in diastole when there is the sudden cessation of rapid ventricular diastolic filling. When accurately recognized, a pericardial knock is a specific but insensitive indicator of pericardial constriction.¹⁻⁷

In many cases of chronic pericardial constriction, definitive treatment is surgical and consists in wide resection of both pericardial layers, however it has a high mortality risk that goes from 6 to 12% according to different case series.^{1,3-9}

CLINICAL CASE

A 41-year-old man with a medical history of pulmonary tuberculosis presented to the emergency department complaining of dyspnea, lower limbs edema and increased abdominal volume of 3 months of evolution. This occasion with exacerbation of symptoms.

On first examination, elevated jugular venous pressure was evident, with an estimated pressure of 11 cmH₂O; rales at the base of both lungs were auscultated, with decreased respiratory sounds at the base of the right lung; generalized edema was present, mainly affecting the lower limbs; first and second heart sounds were diminished in intensity, and a prominent third heart sound (a pericardial knock) in early diastole was present. The patient was admitted to hospitalization for hemodynamic stabilization and further research.

It was documented on electrocardiogram (ECG) the presence of sinus tachycardia of 130

bpm and right bundle branch block with a QRS axis to 90° without any other alteration. Chest X-ray showed right pleural effusion and signs of congestive heart failure with an enlarged cardiac silhouette, redistribution and interstitial edema. It was observed an area of increased density suggestive of a calcified pericardium (Figure 1).

Echo imaging reported dilated right heart cavities with paradoxical septum motion, reduced right ventricular systolic function with a tricuspid annulus plane systolic excursion (TAPSE) of 13 mm (normal > 17), elevated systolic pulmonary artery pressure (SPAP) of 65 mmHg, mild tricuspid regurgitation and a restrictive diastolic filling pattern, with an E/A index of 2.4. The pericardium layers were hyperechoic.

A computed tomography (CT) scan was performed to obtain a better visualization of the pericardium. It showed a thick and heavily calcified pericardium affecting the most part of the cardiac circumference. A moderate right pleural effusion, as documented previously on chest X-ray, was still present (Figure 2 A-C). Extension of the CT scanning to the abdomen, showed hepatic and splenic enlargement, accompanied by moderate ascites (Figure 3).

On his medical history, he referred having been diagnosed of pulmonary tuberculosis

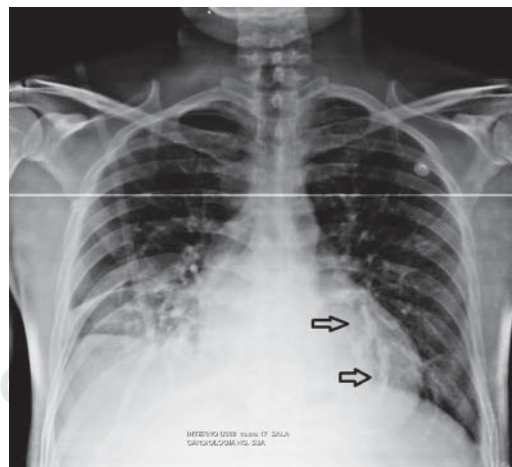


Figure 1. Chest X-ray showing signs of congestive heart failure with right pleural effusion, an enlarged cardiac silhouette, and an image of increased density (arrows) suggestive of pericardial calcium.



Figure 2 A-C. Thoracic CT imaging at different axial segments showing a thickened calcified pericardium and moderate right pleural effusion.



Figure 3. Abdominal CT showing hepatic and splenic enlargement accompanied by ascites.

three years ago and that he interrupted treatment voluntarily at the third month of initiation with no subsequent following up. A skin tuberculin test was performed, being positive.

Calcified tuberculous constrictive pericarditis was diagnosed and first-line antitubercular agents were initiated. Surgical treatment (pericardiectomy) was proposed to the patient, explaining the benefits and risks of the procedure. He refused surgical intervention, and pharmacological therapy for heart failure as per guidelines was implemented.

DISCUSSION

Tuberculous pericarditis is not a frequent cause of heart failure in the developed world, accounting for less than 4% of pericardial disease in these countries; however in underdeveloped countries it does account for more than 90%

of pericardial effusion cases in human immunodeficiency virus (HIV) infected people, and for 50-70% in developing countries where tuberculosis is endemic. Congestive heart failure secondary to chronic compression is the most common clinical presentation. Mortality in these patients is high, ranging from 17-40% at six months after diagnosis.³

In the world extra-pulmonary tuberculosis represents 10% of cases. Tuberculous pericarditis, caused by *Mycobacterium tuberculosis*, is found in approximately 1% of all autopsied cases of tuberculosis and in 1% to 2% of instances of the pulmonary form of the disease. The incidence of the disease is highly variable depending on regional aspects, in Latin America it goes from 0.5% to 1.5% and till 2007 showing an annual decrease of approximately 5%.¹⁰ In 2009 in Mexico 17.5% of tuberculosis cases were extra-pulmonary.¹¹

In México it has been observed an association between tuberculosis and diabetes mellitus, malnutrition, and alcohol and drugs consumption. Overcrowding and a low cultural level are also risk factors to have the disease.¹²

Pericardial involvement usually develops by retrograde lymphatic spread of *M. tuberculosis* from peritracheal, peribronchial, or mediastinal lymph nodes or by hematogenous spread from primary tuberculous infection. The lymphatic drainage of the pericardium is primarily to the anterior and posterior mediastinal and tracheobronchial lymph nodes as suggested by the pattern of lymphadenopathy seen in tuberculous pericarditis.^{13,14}

The immune response of the pericardium to the protein antigens of the bacillus induce a de-

layed hypersensitivity response, stimulating the releasing of cytokines by lymphocytes, which activate macrophages and lead to granuloma formation. This hypersensitivity reaction is orchestrated by TH-1 lymphocytes. The histological pattern seems to be affected by the immune status of the patient, with fewer granuloma formation in HIV-infected patients.¹³

Four pathological stages of tuberculous pericarditis are recognized: (1) fibrinous exudation with initial inflammatory infiltration; (2) serosanguinous effusion with a predominantly lymphocytic exudate with monocytes and foam cells; (3) absorption of effusion with granulomatous caseation and pericardial thickening caused by fibrin and collagen leading to fibrosis; and (4) constrictive scarring.^{13,14}

Nowadays is not common to observe a severe calcification of the pericardium, because it occurs much less commonly in those patients with constrictive pericarditis secondary to radiation or a previous surgery; in which the pericardial thickness may even be normal. However in tuberculous constrictive pericarditis it is not so infrequent to observe calcification of the pericardium.⁴

The main differential diagnosis is made between pericardial constriction and restrictive cardiomyopathy.^{1-9,15} In both conditions ventricular filling is restricted and a high driving pressure across the valves at the time of atrioventricular valve opening results in early rapid diastolic filling and an abrupt increase in ventricular pressure.^{4,7} In a healthy person during inspiration there is an increase in filling of the right ventricle because of enhanced venous return but filling of the left ventricle is unaffected during the whole cardiac cycle. In a patient with constrictive pericarditis, during inspiration the left ventricle is underfilled and there is a reciprocal increase in filling of the right ventricle, while during expiration occurs the opposite.⁴

Constrictive pericarditis is characterized by an early rapid filling and equalization of end diastolic pressures in all four cardiac chambers. Because of increased ventricular interdependence in constrictive pericarditis, the right ventricle expands during inspiration producing a ventricular septal shift towards the left ventricle, without a change in total cardiac

volume. For this reason the expansion of the heart chambers during diastole is limited by the rigid pericardial sac with an impeded atrial contribution to mid- and late diastolic filling, while the early diastolic filling is unimpeded. The predominant ventricular filling will fall in the first third of diastole. This produces the hemodynamic dip-plateau pattern or square root sign of ventricular filling pressures during cardiac catheterization the rapid «Y» descent in the jugular venous pressure and plateau during right heart catheterization.^{4,5,7,16}

Doppler echocardiography is useful for the diagnosis of constrictive pericarditis and help distinguish it from restrictive cardiomyopathy.^{5,7,9} In constrictive pericarditis because of the reciprocal filling changes with respiration, the tricuspid velocity increases in inspirations while the mitral velocity decreases. This phenomenon represents the enhanced ventricular interaction, which characterizes constrictive pericarditis but it is absent in both normal subjects and cases of restrictive cardiomyopathy.⁵

Although pulmonary embolism, right ventricular infarction, and chronic obstructive lung diseases, must be also considered. Several echocardiographic parameters can be used to differentiate among them. Cardiac magnetic resonance (CMR) and computed tomography (CT) are best suited to visualize the pericardium.^{1,3,6-8} In CT and CMR, the healthy pericardium is normally visualized as a fibrous lining surrounding the heart with a minimal fluid layer. Minimal pericardial calcifications are early detectable in thoracic CT.⁵

According to the recent European guidelines transthoracic echocardiography and chest X-ray are recommended in all patients with suspected constrictive pericarditis, though the latter is not useful to visualize calcifications in the pericardium, they can be seen sometimes in lateral views in the posterior border.³ CT and/or CMR are indicated as second-level imaging techniques to assess calcifications, pericardial thickness, and degree and extension of pericardial involvement. However cardiac catheterization is indicated only when non-invasive diagnostic methods do not provide a definite diagnosis of constriction.¹⁻⁸

The benefits of pericardiectomy are well established for permanent constrictive pericardi-

tis. Surgical intervention can provide complete relief of symptoms,⁴ however, a wide resection of both pericardial layers has to be made, and though it can be of great benefit for the affected patients, the mortality risk during the procedure is high, reaching 6-12% according to different reports. Major complications include acute perioperative cardiac insufficiency and ventricular wall rupture.^{3,4,6}

In a case series reported in Mexico 30 years ago, 48% of patients presented complications after pericardiectomy, being low-output heart failure, right atrium lesion, bleeding, left ventricle lesion, pulmonary thromboembolism and pneumothorax in that order the most frequent.¹⁶ In a more recent report from the US in 2004 the most frequent cause of death in the perioperative period was still low-output heart failure.¹⁷

Some patients may not benefit from pericardiectomy and this may be due to myocardial compliance abnormalities, myocardial atrophy after prolonged constriction, residual constriction or other myocardial processes.^{1,5} Techniques have been described in which utilization of laser or ultrasound are effective for the debridement of the adherences of an extensively calcified pericardium to the epicardium, which have an increased bleeding risk. Areas of strong calcification or dense scarring may be left as islands to avoid major bleeding. Occasionally, some patients are candidates to video-assisted thoracoscopic pericardiectomy in experienced centers.^{1,3,6,7}

Long-term survival after pericardiectomy for constrictive pericarditis have been found to be related to underlying etiology. Idiopathic cases have shown the best survival rate than miscellaneous groups, including tuberculous pericarditis. The impact of pericardial calcification on perioperative mortality is still not clear, although some studies have reported an increased mortality in these patients.¹⁷

Medical therapy is supportive, and aimed at controlling symptoms of congestion with diuretics in advanced cases, and when surgery is contraindicated or at high risk.^{5,8}

CONCLUSIONS

Pericardial disease is not a rare form of manifestation of tuberculosis infection, and it usually

have a good response to antitubercular drugs, at least in the first stages of the disease. However severe calcification of the most part of the pericardium, the last stage of progression of tubercular affection, is not frequently observed and has a poor prognosis if not surgically treated.

The case of our patient showed a non-common pattern of severe pericardial affection that played a major role in the therapeutic decisions. As mentioned above the degree of the calcifications and the pericardial adherences to the myocardium posed the patient into a higher risk of mortality during and/or after the surgical intervention, however it is the only intervention that would allow to improve the prognosis in this stage of the disease.

Although there are several risk factors that allow the progression of the disease, our patient lack the majority of them; he had no immunologic compromise, no chronic or degenerative disorders, and referred a low level of alcohol consumption. However his social conditions, a poor understanding of the disease and the importance of treatment adherence, played a major role in leading him to this chronic fatal complication.

ETHICAL DISCLOSURES

- Animal and human protection. The authors declare that in this research no animal or human experiments have been performed.
- Data confidentiality. The authors declare that in this manuscript there are no data that identifies any patient.
- Right to privacy and informed content. The authors declare that in this manuscript there are no data that identifies any patient.

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