Case Report

Postpericardiotomy Syndrome after Nuss Procedure

Síndrome Pós-Pericardiotomia depois de Cirurgia de Nuss

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Introduction

Postpericardiotomy syndrome (PPS) was first described in 1953 in patients with fever and pleuritic chest pain undergoing rheumatic mitral stenosis repair surgery.[1, 2] PPS was initially believed to be associated with rheumatic disease reactivation and was subsequently recognized as an autoimmune inflammatory process.[1, 2]

The proposed diagnostic criteria have changed over time[1] but are currently based on the COPPS[3] and COPPS-2[4] studies, which were developed to assess the benefit of colchicine in PPS.[2] A PPS diagnosis requires at least two of the following criteria: fever of unknown cause, pain with characteristics of pleuritis or pericarditis, a rubbing sound on auscultation, and evidence of pericardial and/or pleural effusion with increased C-reactive protein level.[3,5]

Most patients present a benign and self-limited progression.[6] However, the form and severity of clinical presentation can vary widely from asymptomatic patients with mild pleural and/or pericardial effusion to those with serious complications such as cardiac tamponade.[1, 2]

The objective of this study is to highlight the importance of a high index of suspicion for this clinical entity by presenting the case of a patient with PPS who was diagnosed after a minimally invasive procedure.

Case report

A 17-year-old male adolescent underwent Nuss surgery to correct pectus excavatum. After the procedure, the patient attended regular surgical follow-up consultations. At day 39 (D39) after surgery, he went to the emergency room (ER) with pain in his chest and left shoulder. Findings of a chest X-ray were normal, and he was discharged with analgesia. Due to pain progression, at D58 after surgery, he returned to the ER with a seven-day history of fever, evident prostration, pleuritic pain, and pain on abdominal palpation.

Although hemodynamically stable, an objective examination revealed sweating, prostration, an axillary temperature of 38.6ºC, pain facies, intolerance to supine decubitus, pulmonary auscultation with decreased breath sounds, rhythmic cardiac auscultation with no murmurs, a soft abdomen, depressible and painful on palpation in the right iliac fossa, and no defense but with pain on decompression.

The analytical study highlighted: leukocytes 10,600/μL with 73% neutrophils, platelets 320,000/μL, sedimentation velocity (SV) 121 mm/h, C-reactive protein (CRP) 27.63 mg/dL, and procalcitonin (PCT) 0.16 ng/mL. A chest X-ray showed a slight left pleural effusion with suspected pulmonary consolidation in the lung parenchyma. Empirical antibiotic therapy was defined based on the clinical status of the patient associated with analytical and imaging changes.

Despite treatment, the patient remained prostrate and feverish with localized pain in the left hemithorax and generalized pain on deep abdominal palpation. An abdominal ultrasound showed a slight perihepatic and pelvic hollow peritoneal effusion.

He also underwent electrocardiography (left precordial repolarization disturbance with negative T waves in V4–V6, ST-segment changes on the lower leads, and slight ST-segment elevation in DI and DII) and transthoracic echocardiography, which confirmed the presence of circumferential, heterogeneous, and septate pericardial effusion without diastolic collapse measuring about 29.5 mm in the anterior wall and 38 mm in the posterior wall (videos 1–4). Thus, PPS was the probable hypothesis.

The patient underwent empirical treatment with ibuprofen 600 mg every 8 hours, intravenous methylprednisolone 1 mg/kg/day, and colchicine 1 mg/day. Surgical intervention was chosen to remove the bar. After the procedure, however, the patient's general condition deteriorated, with signs of respiratory distress and absence of breath sounds in the left hemithorax with dullness on percussion. The chest X-ray confirmed a dense opacity of the entire left hemithorax that was treated with pleural effusion drainage. The liquid had a serosanguineous appearance and transudate characteristics (leukocytes 3,590/μL, neutrophils 1,757/μL [49%], glucose 5.0 mmol/L, lactic dehydrogenase 976 IU/L) but no evidence of a bacterial infectious etiology.

The established therapy continued and was changed to oral prednisolone after five days of intravenous steroid therapy. The antibiotic therapy was discontinued when the blood and pleural fluid culture results were negative. Viral serology and autoimmune marker test results were also negative.

Serial echocardiogram evaluations showed progressive resolution of the pericardial effusion with normal systolic/diastolic function but no hemodynamic changes. On D12 after starting therapy, the patient was clinically well, with a SV of 18 mm/h and a CRP level of 0.33 mg/dL, and presented only a thin blade of pericardial effusion (about 5 mm) on echocardiogram. He was discharged with a

Keywords

Postpericardiotomy Syndrome; Diagnosis; Echocardiography.

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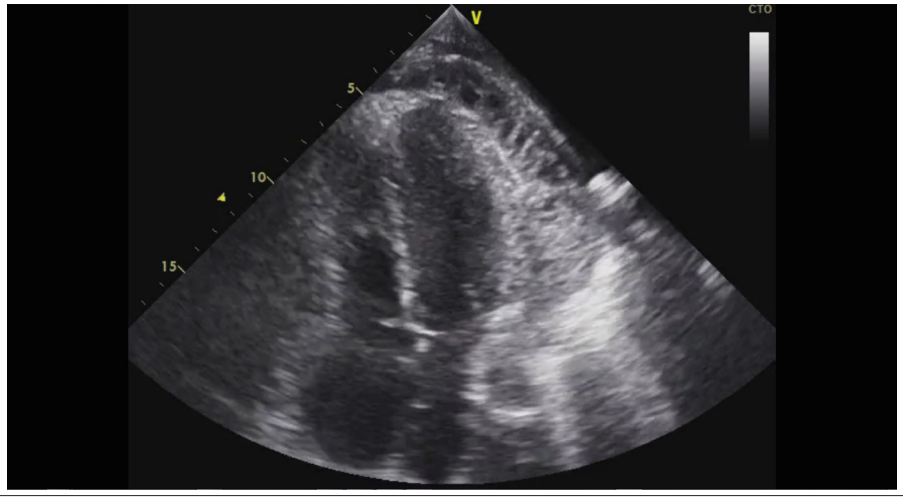
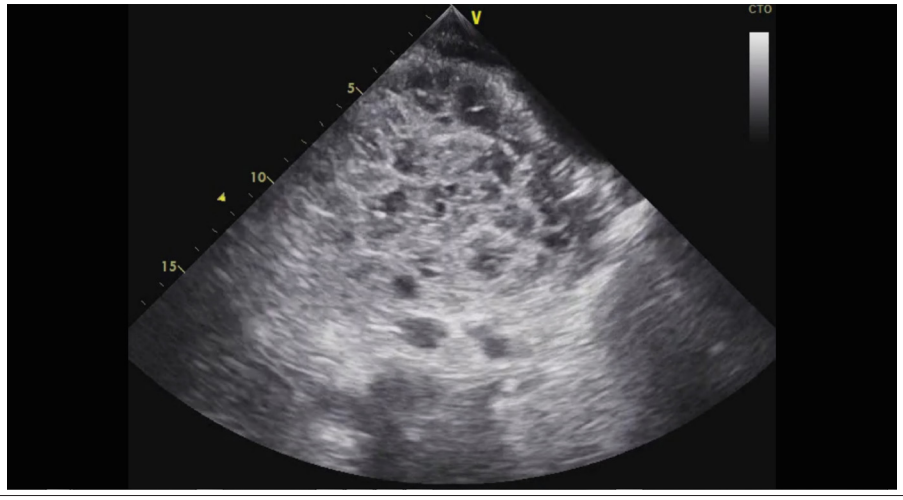
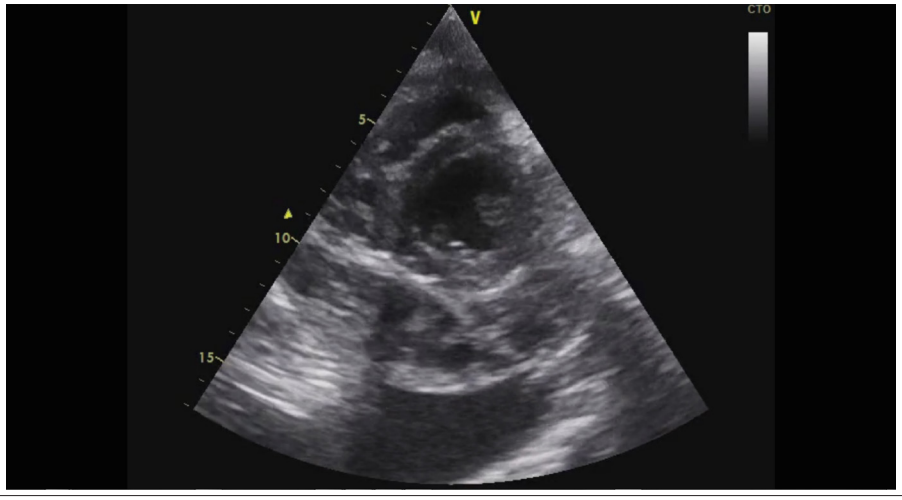


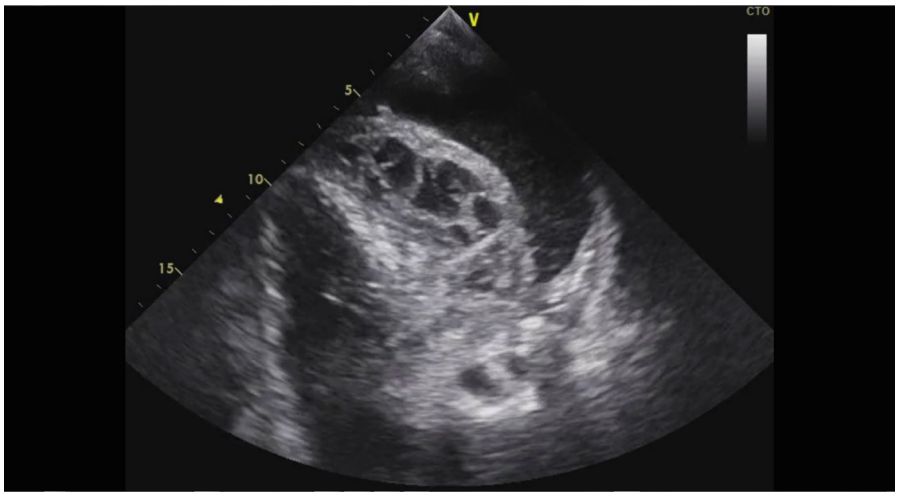
Video 1 – *Transthoracic echocardiogram in apical four-chamber view.*



Video 2 – *Transthoracic echocardiogram in apical view showing heterogeneous and septate circumferential pericardial effusion.*



Video 3 – *Transthoracic echocardiogram in parasternal short-axis view showing septate circumferential pericardial effusion and pleural effusion.*



Video 4 – *Transthoracic echocardiogram in two-chamber apical view showing septate pericardial effusion and pleural effusion.*

prescription for ibuprofen 600 mg every 8 hours, colchicine 1 mg, and a corticosteroid weaning plan. At follow-up visits, the patient continued to be asymptomatic with normal echocardiography findings.

Discussion

PPS is a common complication after cardiac surgery,[3] with an incidence of 10–40% depending on the population studied.[1-4] Although more common in patients undergoing cardiac surgery, PPS is currently associated with a variety of chest wall procedures,[7] even when minimally invasive, such as Nuss surgery,[8] as in the present case. The pathophysiological mechanism underlying PPS, although unknown, is believed to be a result of surgical trauma to the pleura and/or pericardium associated with the procedure,[1,8] with an intraoperative exposure of mesothelial cells and subsequent release of antigens and immune complexes that trigger an autoimmune inflammatory response.[1, 5,7]

The PPS diagnosis is based on clinical criteria[1]; as noticed in this case, it is a real challenge. Symptom onset is quite variable; although they appear more frequently in the first days and/or weeks after the procedure, they can also start after a few months.[1-3] Hence, recognizing this clinical entity is important. In the present case, the first complaints started more than a month after the Nuss surgery in the form of chest pain, initially without other associated symptoms but with a marked worsening of pain and general condition and the onset of fever. The presence of pleuritic pain and fever, associated with evidence of pleural and/or pericardial effusion, constitute clinical data favorable to the diagnosis of PPS, especially when the patient has a previous history of undergoing a procedure potentially associated with this entity, such as Nuss surgery. However, the differential diagnosis of PPS, pleural and pericardial effusion, frequent complications in the immediate postoperative period, is difficult.[5]

Although there are no confirmatory tests[1,7], evidence of an inflammatory process associated with the clinical status is highly suggestive of the disease.[5] Thus, in addition to a complete blood count with leukogram, CRP, and SV,[2,7] PCT plays a fundamental role in the differential diagnosis of infectious diseases. In the case described here, the association of a marked SV and CRP increased with a normal PCT, suggesting a clinical condition resulting from an autoimmune inflammatory response, that is, a non-bacterial etiology. However, at an early stage, considering the patient's clinical status and suspected pneumonia, empirical antibiotic therapy was instituted and maintained until later confirmation on negative culture results.

As it is an autoimmune pathology, autoimmune markers, such as anti-smooth muscle and antinuclear antibodies, were also evaluated but rendered negative results. Since its determination has no influence on the therapeutic decision,[7], these tests may be unnecessary.

As for the therapeutic approach, except in cases with hemodynamic changes requiring an immediate invasive intervention,[9] PPS treatment is empirical[1, 3, 5] and based on drugs with anti-inflammatory activity (nonsteroidal anti-inflammatory drugs, corticosteroids, and, more recently, colchicine), considering the underlying pathophysiological mechanisms. Thus, the therapeutic regimen with ibuprofen and colchicine was instituted and, due to the patient's severe clinical status, corticoid therapy was initiated as recommended in the literature.[5] The bar was also surgically removed. This decision, although controversial, has been described in the literature.[8]

Due to the risk of constrictive pericarditis (about 3% of cases[5]), the patient maintains regular follow-up consultations every 6–12 months with normal assessments to date.

Conclusion

The case described here highlights the need for a high index of suspicion of this entity due to the symptoms that may initially be frustrating and their subacute presentation. Despite an extensive, septate, and heterogeneous pericardial effusion, the patient presented good clinical, laboratory, and

echocardiographic progression under the established therapy, remaining asymptomatic and without residual lesions in subsequent evaluations.

Authors' contributions

Data collection: Lopes L, Henriques C, Francisco A, Rodrigues D, Pires A. Data analysis: Lopes L, Henriques C, Francisco A, Rodrigues D, Pires A. Manuscript writing: Lopes L. Critical review of the manuscript for important intellectual content: Francisco A, Rodrigues D, Pires A.

Conflict of interest

The authors have declared that they have no conflict of interest.

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