Case report

Adult onset Still's disease revealed by a cardiac tamponade: Case report.

- *Abstract:*

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease of unknown origin with various clinical manifestations. We here describe a case of a 57-year-old presenting Still's disease made more difficult by cardiac tamponade.
This case highlights the importance of a prompt diagnosis of cardiac involvement in inflammatory diseases, as it can be fatal, and underlines the interest of echocardiographic evaluation not only in symptomatic patients, but also for the systematic detection of pericardial effusions. In case of hemodynamic compromise, it allows us to determine the timing, approach, and method of pericardial drainage because cardiac tamponade is a very rare complication that requires an invasive approach.

- *Keywords:*

Diagnosis; treatment; cardiac tamponade; adult onset still disease; AOSD.

- *Introduction:*

Still's disease was first described by Still(1) *in 1897 in children, but it was not until the 1970s that Bywaters*(2) *and Bujak et al*(3) *described similar symptomatology in adult patients. Despite the similarity of symptoms, these two entities remained distinct, and the latter was named adult-onset Still's disease (AOSD). It is characterized by daily fever, evanescent rash, arthritis, pharyngitis, leucocytosis, lymphadenopathy, and less frequently, polyserositis and hepatitis. Pericarditis occurs in approximately 30-40% of patients and tamponade remains a scarce complication*(4). *However, the diagnosis remains purely clinical and lacks evidence, hence the need for diagnostic criteria that have been proposed and modified over time.*
We report a case of cardiac tamponade revealing an adult onset Still's disease.

- *Presentation of the case :*

A 57-year-old male resident from a rural area was admitted to our cardiac intensive care unit displaying symptoms of NYHA stage IV dyspnea and retrosternal chest pain, both of which had persisted for 5 days in a febrile state. The patient had a type 2 diabetes, which was being treated with metformin with an unknown degenerative profile. It is also noteworthy that the patient had a 2-year history of untreated polyarthralgia with an inflammatory pattern and, on presentation, had salmon coloured rash on his chest and abdomen. The patient's infectious history was determined to be negative upon inquiry.

1- Clinical findings:

Upon initial evaluation, the patient presented with acute dyspnea and had an oxygen saturation level of 95% while breathing room air. Vital signs showed a blood pressure of 100/60mmHg, a regular heart rate of 110bpm, and paradoxical pulse. The examination also revealed signs of right heart failure, including spontaneous jugular vein distention and pitting edema in the lower extremities. The thoracic examination indicated dullness in the bases and muffled heart sounds, with no murmurs or rubs detected. Despite the patient high fever at 39°, its infectious history was determined to be negative upon review with no clinical sign suggesting arthritis.

2- Diagnostic assessment and therapeutic intervention:

Initial laboratory data revealed a leucocytosis of 16 220 with 92% segmented neutrophils, hemoglobin of 16.22g/dL, elevated inflammatory markers; erythrocyte sedimentation rate [ESR] at 97, C-reactive protein [CRP] at 255 and fibrinogen at 7, procalcitonin was negative at 0.4 and ferritin level was increased at 1322 with a glycosylated ferritin fraction (GF) low at 12.4%, an empiric antibiotic therapy was initiated for a presumed infectious etiology, however fevers and leucocytosis persisted while on therapy. A thorough infectious investigation was unrevealing, including blood cultures, fungal cultures and serologies. A chest X-ray revealed marked cardiomegaly (CTI 0.8) with minor bilateral pleural effusion. The electrocardiogram (ECG) demonstrated sinus tachycardia and diffuse microvoltage. Shortly thereafter, the patient presented a hemodynamic distress as his BP dropped to 85/54. An emergency bedside transthoracic echocardiography (TTE) was performed and revealed a large pericardial effusion , with abnormal respiratory variation in transvalvular blood flow velocities, a paradoxical septum on inspiration, and a significant dilatation of the inferior vena cava, all consistent with the diagnosis of tamponade. In accordance with our triage strategy, an urgent pericardiocentesis was performed, large amount of haemorrhagic fluid was drained. The patient's distress abated following the pericardiocentesis and pericardial fluid was exudates, it yielded negative microbiologic examination including cytology and cultures (aerobic, anaerobic, acid-fast and fungal) and was unremarkable upon histologic analysis. An immunologic screening for rheumatoid factor and anti-nuclear antibodies (ANA) was drawn and was also negative. The patient received aspirin 3g/d during the investigation phase with no clinical nor biological improvement.

Once the diagnosis of AOSD is established based on the presence of fever, arthralgia, skin rash and leucocytosis clinically and hyperferritinemia with a decreased glycosylated fraction, the patient's antibiotics were discontinued once cultures were finalized as being negative, then corticosteroids were required to induce symptom remission starting with intravenous infusion of high-dose methylprednisolone for 3 days in a row then it was tapered down to prednisolone oral route (1 mg/kg/d) with a remarkable clinical et biological improvement during the first week with a resolution of fever and the return of leucocytosis. after a rapid prednisolone tapering scheme, the patient was then discharged and was kept on a regimen of prednisone 20mg/d.

3- Follow-up and outcomes:

(During first months of follow-up, patient was asymptomatic and TTE revealed a complete resolution of pericardial effusion. Eight months after hospitalization, the patient continued to do well, at which time he was weaned off steroids. Approximately a year after hospitalization, he developed recurrence of arthralgias for which he was restarted on maintenance doses of steroids.)

A close follow up was maintained during the initial months following hospitalization, and it was determined through a transthoracic echocardiogram (TTE) that there was a complete disappearance of the pericardial effusion as well as the pleural effusion, with the patient remaining asymptomatic throughout this time period.

Eight months after the initial hospitalization, the patient continued to exhibit excellent health, and it was deemed appropriate to begin a gradual weaning process off of the steroid medication. Despite this positive progression, approximately one year after the initial hospitalization, the patient experienced a recurrence of arthralgias as he was weaned off prednisone.

In light of this development, it was necessary to restart the patient on a maintenance dose of prednisone in order to manage the symptoms and maintain the overall well-being.

- *Discussion:*

1. *Estimates indicate that the incidence of AOSD in the general population falls within a range of 0.16 to 1.47 per 100,000 individuals. Among patients with AOSD, fluid in the pericardial sac has been identified in 30 to 40% of cases. However, it should be noted that not all individuals with AOSD will exhibit symptoms of pericarditis.(5)*
2. *The onset of Still's disease can be sudden and is characterized by a triad of symptoms, including a high fever accompanied by a transient salmon-colored rash appearing at fever peaks and arthralgia.(2) The diagnosis is established on a constellation of clinical manifestations, laboratory findings, and exclusion of other systemic infections, conditions, and malignancies. Despite the lack of a specific diagnostic test for Still's disease, several authors have attempted to develop criteria that aid in its diagnosis based on observations of various patient groups.(5) Many diagnostic criteria were propose, however, the criteria proposed by Yamaguchi et al. being the best-known and characterized by a high sensitivity of 93.5%.(6) The constellation of clinical manifestations and laboratory findings in our case satisfied the diagnostic criteria of AOSD proposed by Yamaguchi et al. (6)*

3. *Serosal involvement is seen in 25% to 60% of all AOSD patients, and cardiac involvement is frequently observed and can be severe. Pericarditis is present in 10% to 40% of patients, with 20% of these patients experiencing pericardial effusion or cardiac tamponade.*(7)
   Pericarditis is sometimes discovered fortuitously in a patient diagnosed with AOSD on a routine EKG, chest x-ray, or TTE, it is often accompanied by a pleural effusion in 60 to 80% of cases, and presents during the initial flare-up of the disease. Pericarditis or its complications can be the first symptom of AOSD, however, this does not appear to negatively impact the prognosis. Myocarditis is less common, affecting about 3% of patients, and can lead to various complications such as complete atrioventricular block, tachyarrhythmia, heart failure, or cardiogenic shock. Endocardial involvement is uncommon, and can manifest as non-infectious endocarditis.(7)
   The occurrence of cardiac tamponade is exceptional, especially during a flare-up of AOSD, and only few cases were reported. When tamponade reveals the disease, it can make diagnosis difficult, as other symptoms may be overlooked. If patient presents signs of tamponade, pericardial drainage may be required, either through an ultrasound-guided puncture or a sub-xiphoid surgical procedure. However, treatment with high doses of corticosteroids, often in the form of methylprednisolone, has been successful when the diagnosis of AOSD is suspected with no hemodynamic distress. (8)
   In AOSD, the pericardial fluid is commonly sero-hematous and exudative. While pericardial biopsies are rarely performed, they often reveal either an acute, non-specific pericarditis or a chronic, oedematous pericarditis. Congestive heart failure has been reported in few cases, and limited endomyocardial biopsies have shown only diffuse, non-specific mono-nuclear inflammation, along with fibroblast proliferation and hypertrophy in certain myocardial fibers. However, endocardial lesions are uncommon in this disease. In rare cases, mitral or aortic valve involvement has been reported, requiring valve replacement in some patients.(9)
   Due to the frequency and severity of cardiac involvement in AOSD, it is recommended that all patients undergo regular echocardiographic evaluation to monitor for these complications
4. *The primary line of treatment for patients with symptoms that warrant medication typically involves the use of nonsteroidal anti-inflammatory drugs (NSAIDs). When necessary, other options such as corticosteroids, immunosuppressive medications like methotrexate or cyclophosphamide, or biologic drugs such as tumor necrosis factor inhibitors, interleukin-1 receptor antagonists, and anti-B-cell antibodies may be prescribed.*(10) *In general, cardiac involvement tends to respond well to conservative therapies, but in cases of cardiac tamponade, percutaneous or surgical pericardial drainage may be required.* (4)*The prognosis for adult-onset Still's disease is generally favorable, with low mortality rates and a 10% annual recurrence rate during follow-up.*(11)

5. *This case highlights the importance of considering a broad differential diagnosis for patients presenting with cardiac tamponade, including rare conditions like adult onset still disease. Additionally, this case underscores the need for a thorough medical evaluation to uncover underlying causes, even in the absence of characteristic symptoms. This is particularly important for patients with pre-existing medical conditions, such as diabetes, who may be at an increased risk for developing secondary conditions.*

- *Patient perspective and informed consent:*

- *Conclusion:*

This case report re-emphasizes the importance of considering Still disease in the differential diagnosis of the life-threatening emergency of pericardial tamponade. AOSD often poses a diagnostic and therapeutic challenge and clinical guidelines are

lacking. The emergence of validated diagnostic criteria, discovery of better serological markers, and the application of new biological agents may all provide the clinician with significant tools for the diagnosis and management of this complex systemic disorder.

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