

Sweet's Syndrome with Severe Cardiac Involvement

Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, is a rare skin condition marked by the sudden appearance of erythematous and painful papules, plaques or nodules, mainly on the arms, neck, face and back, associated with fever, leukocytosis, neutrophilia and diffuse neutrophil infiltrate. (1)

The pathogenesis is not clearly established and is described as multifactorial. (2)

There are three presentation forms, including the idiopathic or classic, associated with oncologic or onco-hematologic diseases as the paraneoplastic syndrome and the drug-induced form (especially associated with granulocyte colony stimulating factor). (1)

Systemic involvement is extremely rare. There are few case reports in the literature that describe this syndrome with cardiovascular involvement. (1, 3, 4)

We present the case report of a patient with confirmed Sweet's syndrome, with severe pericardial involvement and hemodynamic decompensation.

He is a 42 year-old man without any relevant cardiovascular history, diagnosed with Sweet's syndrome confirmed by a skin biopsy a year ago. During follow-up, a myelodysplastic syndrome in leukemic phase was discovered, currently being treated with azacitidine, and in the waiting list for bone marrow allogeneic transplantation

Our patient had a prolonged hospital stay because of febrile neutropenia with possible epididymo-orchitis, for which he completed antibiotic therapy without germ identification in cultures.

During hospitalization he presented with fever and new skin lesions compatible with Sweet's syndrome reactivation. This was associated with neutrophil increase due to granulocyte colony stimulating factor administered a few days before.

Concomitantly, he presented with pericardial-type thoracic pain, accompanied by shortness of breath in functional class IV, with clinical parameters of shock. Laboratory testing revealed pancytopenia, normal renal function and normal cardiac markers, including ultra sensitive troponin. NT-proBNP level was 14,000 pg/ml. The chest x-ray showed new onset cardiomegaly without flow redistribution (Figure 1). Electrocardiogram showed sinus tachycardia, low voltage in peripheral leads and diffuse ST-segment elevation of 1 mm (Figure 2).

Transthoracic echocardiogram revealed new severe left ventricular systolic dysfunction involving apical segments that was not present on the last echocardiogram performed 14 days before, without significant valve diseases or pericardial effusion.

A Swan-Ganz catheter was introduced showing a mixed distributive or cardiogenic pattern.

Non-invasive ventilation and intravenous diuret-

ics were started, with good response and without requiring the use of inotropes.

With presumptive diagnosis of myopericardial involvement in Sweet's syndrome, hydrocortisone 1 mg/kg/day was started. At 48 hours, clinical parameters, invasive monitoring values and ventricular function were normalized.

A myocardial biopsy was performed that showed the typical interstitial edema and perivascular neutrophil infiltrate, confirming Sweet's syndrome diagnosis.

Sweet's syndrome is characterized by sudden fever, neutrophilia and skin lesions associated with neutrophil infiltrates. Systemic involvement is rare. (5) Cohen et al. described extracutaneous manifestations in Sweet's syndrome associated with hematologic diseases, especially acute leukemia and myelodysplastic syndrome. (2) Central nervous system, bone, lung or

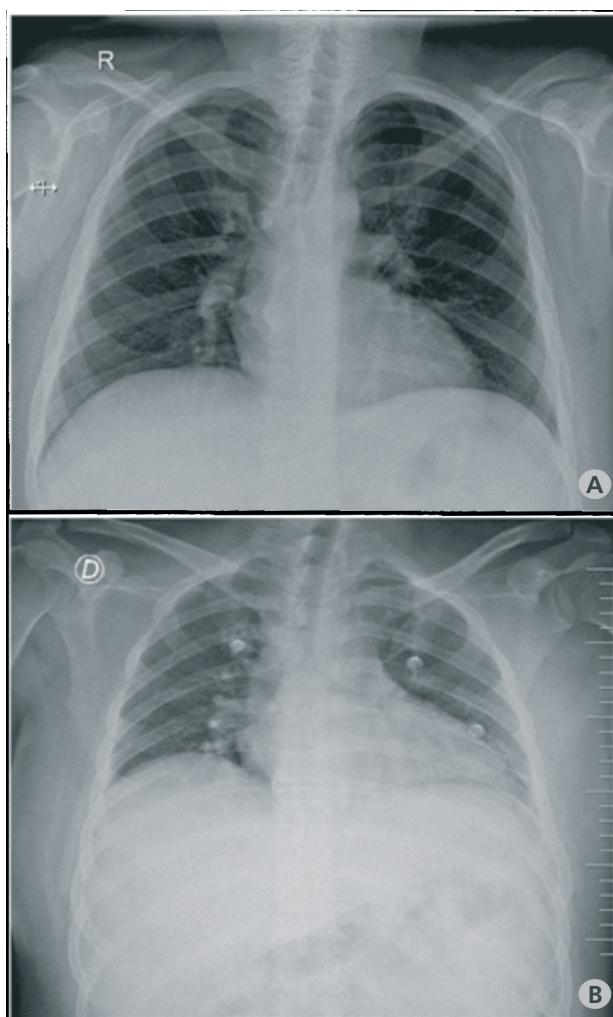


Fig. 1. Chest x-ray: **A.** Normal chest radiography 48 hours prior to decompensation. **B.** Acute phase with shock symptoms and new sudden cardiomegaly.

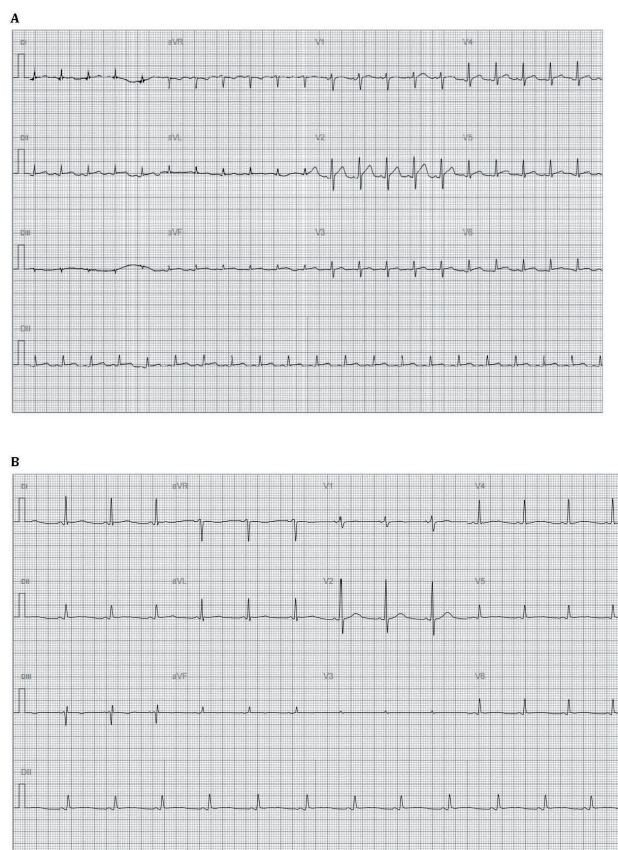


Fig. 2. Electrocardiogram: A. Acute phase: Sinus tachycardia, low voltage in peripheral leads and diffuse ST-segment elevation of 1 mm. B. Recovery phase: Voltage and ST-segment normalization 48 hours after high-dose corticosteroid administration.

hematologic involvement is widely described in the literature, (2) but cardiac manifestations are poorly reported.

This syndrome presents as a multifactorial pathology, generating a systemic inflammatory state that simulates septic shock, with evidence of cytokine release. (3) Our patient was receiving complete antibiotic treatment, without any germ identification in cultures, presenting with severe deterioration of systolic ventricular function associated with parameters of inflammation in the invasive monitoring. This coincides with neutrophil increase secondary to administration of granulocyte colony stimulating factor. Few reports in the literature describe drug induced Sweet's syndrome in patients treated with this medication.

Pathological findings in the myocardial biopsy were consistent with neutrophil perivascular and interstitial infiltration, consistent with Sweet's syndrome (Figure 3). (4)

High-dose systemic corticosteroid administration represents the gold standard treatment for Sweet's syndrome, and in this particular case, our treatment, as well as intravenous diuretics and non-invasive ventilation, was enough to have full hemodynamic and left ventricular systolic function recovery. (2-4)

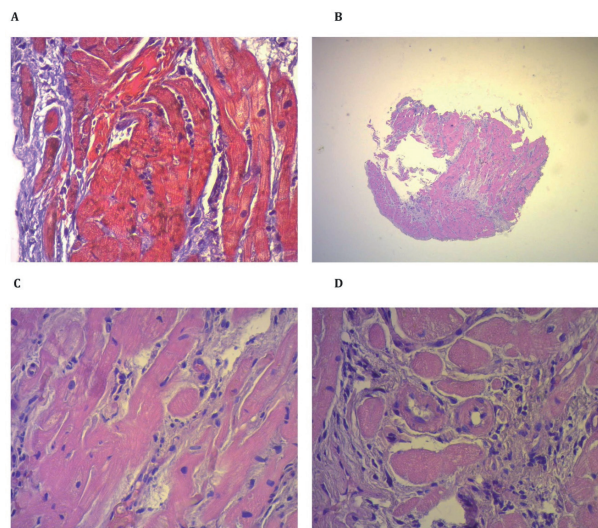


Fig. 3. Pathological findings: Interstitial and perivascular neutrophil infiltrate, with interstitial edema. A. Masson's trichrome stain. B, C & D. Hematoxylin and eosin stain.

As shown in this case report of our patient, early diagnosis of this syndrome and immediate high-dose corticosteroid therapy, together with heart failure treatment, is a cornerstone in the patient's full recovery.

Data from larger number of patients with this syndrome and cardiovascular involvement are needed in order to properly assess the effect of early corticosteroid administration in mortality reduction and complete left ventricular systolic function recovery.

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Primary Cardiac Lymphoma

Primary cardiac lymphoma (PCL) is a non-Hodgkin's lymphoma that occurs only in the heart. It is a very rare condition and is reported to be increasingly diagnosed pre-mortem. We present the case of an 87 year-old woman referred with a two-month history of dyspnea and peripheral edema. The transthoracic echocardiogram (TTE) and the computed tomography (CT) of the chest revealed an 8 cm right atrial (RA) mass. The tumor was excised, and the patient had a favourable immediate postoperative outcome.

The patient, with a 2-month history of shortness of breath worsening and increased leg edema was referred from a local hospital with diagnostic images suggesting a RA mass. Clinical examination, except for the peripheral edema, was unremarkable. Blood screening tests showed a GFR of 35 ml/min. The TTE revealed a large RA mass not resembling a thrombus and without extension into the inferior vena cava (IVC). This was confirmed by a CT scan, showing that the mass was 8 cm in size and had a broad attachment extending from the roof of the RA to the interatrial septum (IAS) (Figure 1). There was no other intrathoracic or abdominal pathology.

Despite her age, surgical removal of the tumor was justified, due to its obstructive nature and the symptoms it caused.

During surgery, the tumor did not appear to be a myxoma. Full resection was not possible due to invasion of the RA wall. The IAS was resected and closed with a CorMatrix patch. During the first few hours after the operation, the patient developed sudden

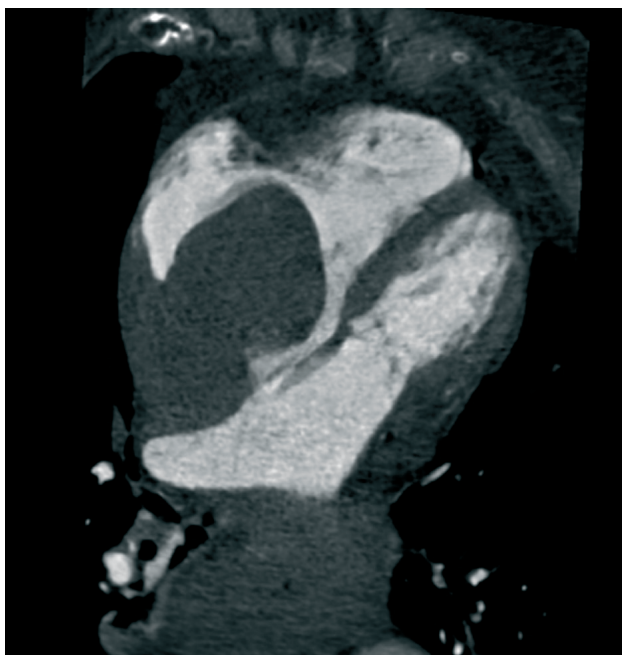


Fig. 1. Image confirming cardiac tumoral mass (see text for description).

ventricular fibrillation and cardiac arrest, requiring a new urgent sternotomy. There was no evidence of tamponade or bleeding, and after internal cardiac massage and a single internal defibrillation, the heart recovered sinus rhythm.

No cause for this event could be found. The patient made a satisfactory recovery, with only acute kidney injury but maintaining good diuresis. She was transferred to her local hospital on the 13th postoperative day, for the oncology team to decide on further treatment.

Histology results indicated a diffuse type B cell lymphoma.

Primary cardiac lymphomas are rare. The rate of occurrence is less than 1.5% of all primary cardiac tumors. The most common clinical presentations are heart failure, pericardial effusion and atrioventricular block. The histological type in immunocompetent patients is diffuse large B cell-type in more than 80% of cases. In immunocompromised patients there is a high chance of much more aggressive cell types.

In general, the prognosis is poor, although patients are very sensitive to chemotherapy. Median survival is 12 months. The preferred localization is the right atrial or ventricular wall (92% of cases), followed by the left atrium and ventricle.

The diagnostic approach is made by TTE, magnetic resonance and CT scan of the chest, abdomen and pelvis. CT scan is particularly useful to exclude other sites of disease.

The main differential diagnoses are myxoma, other cardiac tumors, or thrombus formation.

Surgery is usually indicated in the presence of an intracardiac tumor, although resection is usually incomplete due to the invasive characteristic of the PCL.

Once the diagnosis is confirmed by histopathology, chemotherapy is the best treatment option. Cases of RA wall perforation have been reported caused by tumor lysis syndrome after chemotherapy.

Primary cardiac lymphomas are very rare, but they are currently diagnosed more frequently pre-mortem. Absence of extracardiac diseases is associated with moderate survival improvement, but overall, prognosis is very poor.

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Secondary Cardiac Plasmacytoma

Malignant cardiac tumors are rare and, among them, cardiac plasmacytoma is extremely uncommon.

We present the case of a 68-year-old woman with multiple myeloma (MM) under chemotherapy treatment, who was admitted to the emergency department due to a 3-month history of episodes of dyspnea and symptoms suggestive of a heart condition. A PET-CT scan had previously revealed multiple hypermetabolic masses mainly in the right chambers, consistent with an active cancer process. A magnetic resonance imaging (MRI) was performed for better anatomical mapping of the interference of these masses with cardiac motility.

In addition, the purpose of this work is to present a brief update of this uncommon heart condition, and to highlight the usefulness of imaging methods, especially PET-CT scan and MRI, which have been shown to be valuable for proper diagnosis and follow-up.

The patient consulted for a 3-month history of dyspnea. A control PET-CT scan revealed multiple hypermetabolic lesions located mainly in the heart and affecting the right adrenal gland, the right pelvic cavity, and the abdominal wall (with no evidence of bone hypermetabolic lesions), consistent with an active cancer process (Figure 1). In view of this condition, MRI was suggested for better anatomical discrimination, and to study the cardiac dynamics affected by these masses.

The study was performed using a Siemens Avanto 1.5T system (Siemens, Erlangen, Germany) with surface coils and ECG gating. All the images were obtained during a simple apnea. Anatomical spin-echo, STIR sequences and functional SSFP (True FISP) cine sequences were performed in 2, 4 and 3 chamber cardiac axes, and short-axis sequence from base to apex.

Image analysis showed multiple isointense, pedunculated masses with respect to the myocardium in SSFP cine-MRI and T1 sequences, behaving as hyperintense in STIR sequences.

Two pedunculated masses were identified in the interventricular septum, one of them prolapsing towards the left ventricular outflow tract during systole. Two images in the right atrium were also observed, one of them implanted in the tricuspid annulus and prolapsing towards the ventricular chamber.

STIR images (Figure 2) showed hyperintense fo-

cal areas in mid inferior, mid inferolateral, and basal anteroseptal segments, which would be associated to myocardial infiltration areas due to the tumoral process. Despite these findings, biventricular systolic function was preserved.

Diagnosis was made through leukemia and lymphoma immunophenotyping of the cardiac masses by flow cytometry, revealing a cytometric profile consistent with moderate size LNH B CD10+. These findings were consistent with plasma cells and suggestive of plasmacytoma.

Multiple myeloma (MM) is characterized by uncontrolled proliferation of plasma cells, generally restricted to the bone marrow. (1) Extramedullary MM spread occurs when malignant plasma cells form tumors (plasmacytomas) in other parts of the body outside the bone marrow, whose reported incidence in newly diagnosed MM ranges from 7% to 18% of patients, or 6% to 20% of cases during the course of the disease. (1-3)

The mechanisms of extramedullary spread in MM are poorly understood. Extramedullary spread in MM can have two different origins: direct extension from skeletal tumors when they disrupt the cortical bone; or hematogenous metastasis to distal organs. However, scientific evidence is not enough to demonstrate it. (1)

Extramedullary masses may arise as primary plasmacytomas in patients with normal bone marrow analysis, or manifest themselves in patients with confirmed diagnosis of MM (secondary plasmacytoma), the latter being more aggressive. (4-6)

According to published series, 80 to 85% of patients with extramedullary MM had plasmacytomas in muscles, tendons, fat, or digestive tract, and 15% of the cases in glands, liver, kidneys, respiratory airway, skin, or breast; cardiovascular system involvement was extremely rare or absent. (4, 5, 7) In a review of the literature, Keung et al. found 9 cases of extramedullary MM involving the heart followed by a report

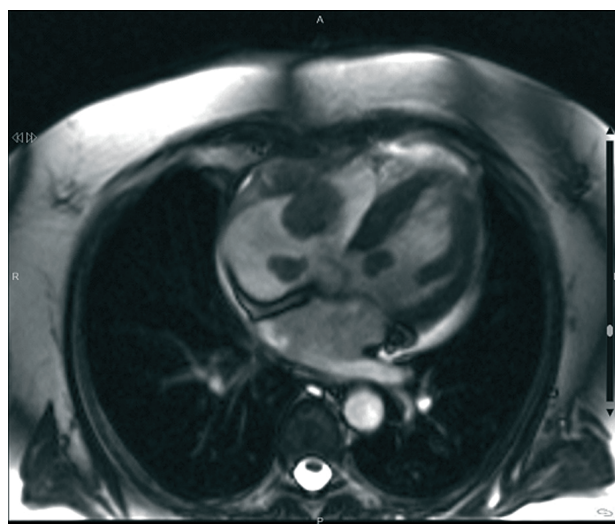


Fig. 1. See text for description.

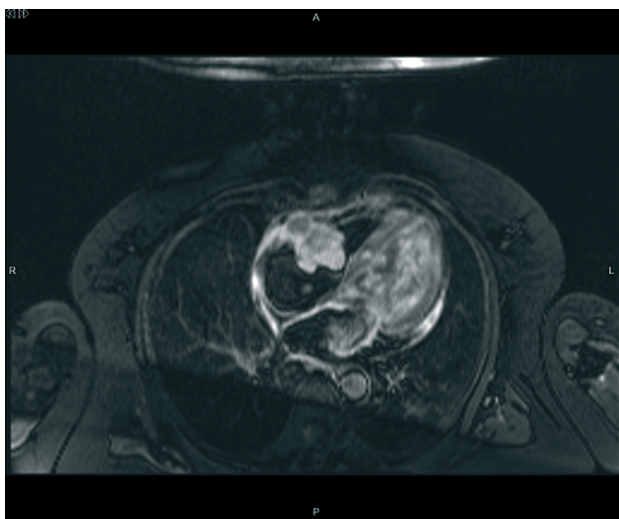


Fig. 2. See text for description.

of a case presenting as a cardiac emergency that required surgical intervention. (8)

The differential diagnosis should be made with all the cardiac masses involving the right heart, particularly metastases.

The extensive use of increasingly sensitive imaging techniques, such as CT scan, MRI, and PET-CT scan, have lately increased the diagnosed cases of extramedullary MM. (1, 7) MRI offers a full range of tools to localize, characterize and evaluate cardiac masses and their physiological effects. In addition, it allows studying the myocardial condition, establishing whether the myocardium has been infiltrated by the neoplasm in question. On the other hand, the PET-CT scan shows involvement at different levels, providing additional information that can be crucial for the treatment and follow-up of this condition. (6) The case we have reported demonstrates the usefulness of these techniques.

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