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 was14,000 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 diuretics 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

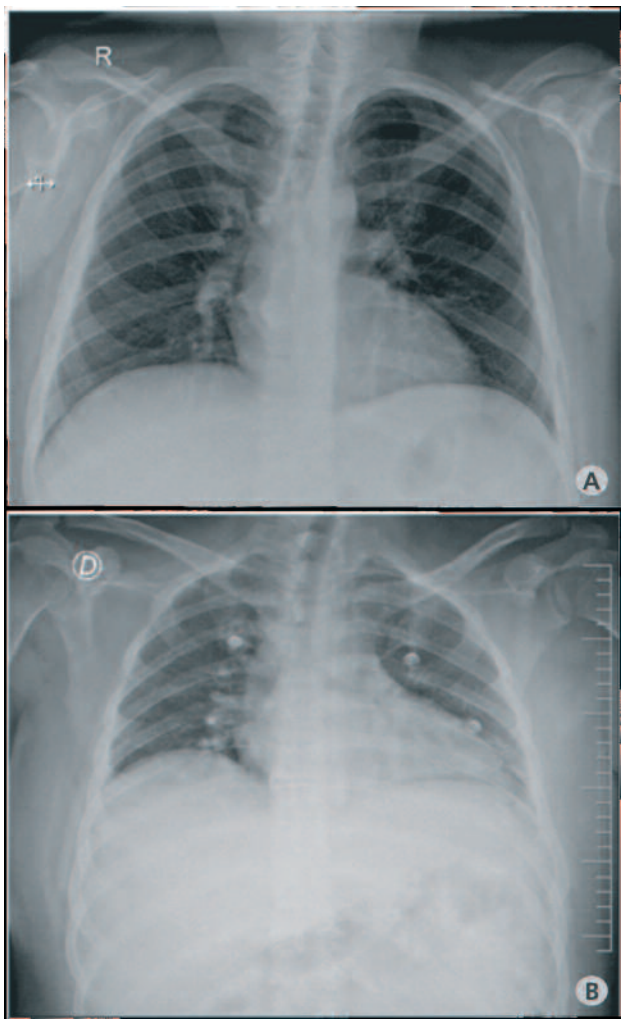

Fig. 1. Chest x-ray: **A.** Normal chest radiography 48 hours prior to decompensation. **B.** Acute phase with shock symptoms and new sudden cardiomegaly.

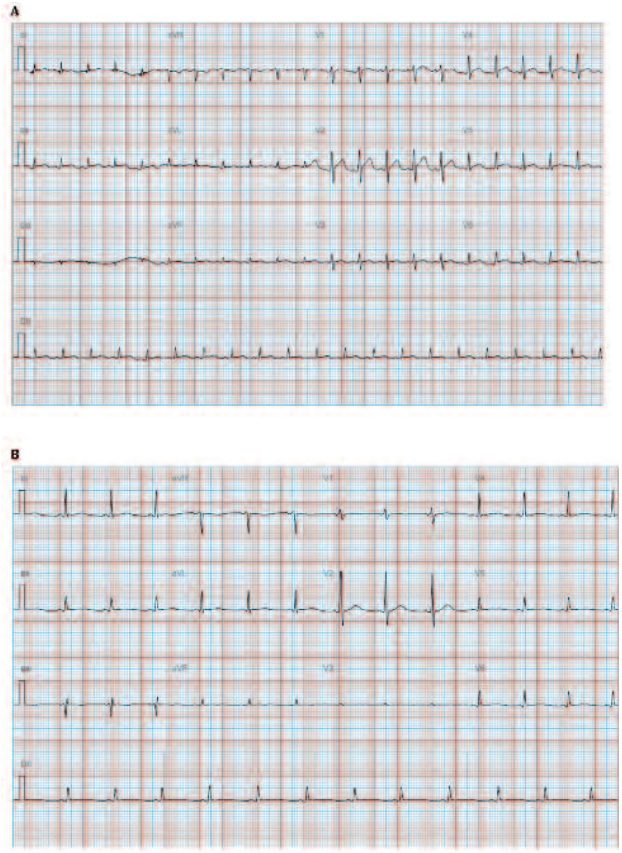

Fig. 2. Electrocardiogram: A. Acute phase: Sinus tachycardia, low voltage in peripheral leads and diffuse ST-segment elevation of 1 mm. B. Recovery phase: Voltage and ST-segment normalization 48 hours after high-dose corticosteroid administration.

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)

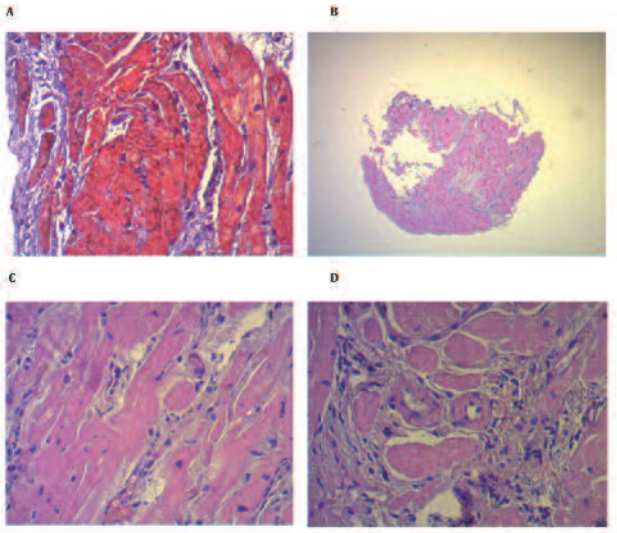

Fig. 3. Pathological findings: Interstitial and perivascular neutrophil infiltrate, with interstitial edema. A. Masson's trichrome stain. B, C & D. Hematoxylin and eosin stain.

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 asses the effect of early corticosteroid administration in mortality reduction and complete left ventricular systolic function recovery.

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REFERENCES

1. Cohen PR. Sweet's syndrome: a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007;2:34. http://doi.org/dvtdb5
2. Raza S, Kirkland RS, Patel A, Shortridge JR, Freter C. Insight into Sweet's syndrome and associated-malignancy: A review of the current literature. Int J Oncol 2013;42:1516-22. http://doi.org/bdhf
3. Kassardjian M, Holland V, Leong T, Horowitz D, Hirokane J. Sweet's syndrome presenting in concordance with acute coronary syndrome. J Clin Aesthetic Dermatol 2012;5:47-5.
4. Shimizu K. Neutrophilic infiltration of the myocardium in a patient with myelodysplastic syndrome. Am J Hematol 1998;58:337-8. http://doi.org/ct3x4g
5. Delabie J, De Wolf-Peeters C, Mooren M, Karien K, Roskams TDV. Histiocytes in Sweet's syndrome. Br J Dermatol 1991;124:348-53. http://doi.org/b3378x

Rev Argent Cardiol 2016;84:247-248. http://dx.doi.org/10.7775/rac.v84.i3.8054