Neutrophilic dermatosis of the hands (Sweet syndrome of the hands) associated with rheumatoid arthritis

Dermatosis neutrofílica de las manos (Sweet de las manos) asociada a artritis reumatoide

Dear Editor,

The predominant involvement of the hands in the Sweet syndrome (SS), pyoderma gangrenosum or pustular vasculitis is rare within the spectrum of neutrophilic dermatosis and is often associated with a concealed neoplasm or another systemic inflammation. Neutrophilic dermatosis of the hands may be medically indistinguishable from other entities but histologically they may differ. However, controversy still abounds today with regard to whether this entity would be a localized variant of SS or an entity in itself.1,2 We present the case of a 77-year-old patient, with a history of diabetes mellitus and high blood pressure. He had been suffering for eight months with painful skin lesions on his hands. The lesions were infiltrated, erythematous violet-coloured plaques, some areas with surface flaking and some areas with a necrotic centre. Due to joint pain and swelling in the hands of over 6 months evolution the patient was referred to the rheumatology department where through clinical and ultrasound examination (with double scan power Doppler synovitis being observed) bilateral arthritis with involvement of the phalangeal metacarpus and right carpus were substantiated. Analysis showed ESR of 28 mm the first hour and a PCR of 61.34 mg/l. The rheumatoid factor was positive (182 U/ml) and the other autoimmune tests were negative. Two skin biopsies were performed where a thick infiltration of neutrophils was observed, with no vasculitis, confirming diagnosis of neutrophilic dermatosis. Cultures were negative. Diagnosis of rheumatoid arthritis was confirmed by the patient meeting with the 2010 classification criteria (8 points).

Topical treatment with clobetasol propionate was initiated with slight improvement and later with methylprednisolone at a 4 mg/day dose, with resolution of the skin lesions and clinical improvement of the arthritis. Treatment with NSAIDS was not initiated because the patient was pending surgery for a knee replacement due to a degenerative disease.

In the year 2000, Galaria et al.3 suggested the term “neutrophilic dermatosis of the hands” as an entity with this single clinical presentation, emphasizing the importance of carrying out differential diagnosis to an infectious process, or another vasculitic coagulopathy or neoplasm process. Neutrophilic dermatosis of the hands or acral may be clinically indistinguishable from SS, but is differentiated from it because it does not meet with the additional minor criteria for diagnosis (fever, previous upper respiratory tract infection, association with malignancy and changes in lab test results).4 Once diagnosis has been confirmed, associated disease has to be ruled out, mainly an intestinal inflammatory disease or a neoplasm. In one published series5 of 36 cases of neutrophilic dermatosis of the hands the age at diagnosis was 61 years, with the female sex predominating and the most common diseases associated with it were myelodysplasia/pre leukaemia (25%), ulcerative colitis (14%), Crohn’s disease (6%), seropositive arthritis (6%) and carcinoma (6%). A case of neutrophilic dermatosis was recently published associated with a primary immune thrombocytopenia, with good response to corticosteroids.6

We believe we have reported a new case of neutrophilic dermatosis of the hands associated with rheumatoid arthritis, the treatment of which with oral corticosteroids improved the skin condition.

References


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