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Neutrophilic dermatosis of the dorsal hands in a patient with polycythemia vera

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To the Editor,

Neutrophilic dermatosis of the dorsal hands (NDDH), a term coined in 2000 by Galaria et al., is an entity clinically and histopathologically similar to Sweet's syndrome (acute febrile neutrophilic dermatosis) (1, 2). The first authors to describe this condition were Strutton et al. (1995), who described six patients with an eruption limited almost entirely to the dorsal hands clinically and histologically resembling Sweet's syndrome. Because of its rarity, its pathophysiology is poorly understood. Although it has many aspects in common with Sweet's syndrome (namely, the fact that histologically there is a dense neutrophilic infiltration of the upper dermis in the lesions), some particular characteristics, especially its confined anatomical location, help differentiate NDDH from the classic form of acute febrile neutrophilic dermatosis (3).

We report the case of a 73-year-old Caucasian man with a diagnosis of polycythemia vera 8 years earlier, initially treated with hydroxyurea, that had been stable for the past 6 years with phlebotomy treatment every 6 months. The diagnostic criteria for polycythemia vera adopted were those of the World Health Organization. Our patient had raised hemoglobin, splenomegaly, no secondary erythrocytosis, and no elevated erythropoietin (16.5 mIU/ml, normal range 3.7–31.5 mIU/ml). He presented with acute nonpruritic, non-scaling, erythematous tumid papules and plaques on the dorsal aspects of both hands (Figs. 1a, 1b). The patient had no systemic manifestation including fever, arthralgia, or general malaise. Blood analysis revealed an ESR of 71 mm / 1st hour (normal range < 30 mm), an erythrocyte count of 5.75 × 10^{12}/L (normal range 4.5–5.9 x10^{12}/L), hemoglobin 17.8 g/dL (normal range 13–17.5 g/dL), hematocrit 52% (normal range 40–50%), leukocyte count 12.1 × 10^9/L (normal range 4.0–11.0 × 10^9/L), neutrophil count 6.39 × 10^9/L (normal range 1.9–7.5 × 10^9/L), eosinophil count 0.28 × 10^9/L (normal range < 0.5 x10^9/L), and platelet count 194 × 10^9/L (normal range 150–450 × 10^9/L). Electrolytes, renal and liver function panel, and urinalysis were normal. A skin biopsy was done, and histopathological examination revealed dense neutrophilic infiltrate of the dermis, mostly perivascular, with some eosinophils and without signs of vasculitis (Figs. 2a, 2b). Based on the clinical, histopathological, and laboratory data, a diagnosis of NDDH was considered. The patient was treated with betamethasone dipropionate cream bid for 6 weeks with progressive resolution of the lesions. After 4 months of follow-up, the patient had no relapse of the skin lesions, presenting only slight residual post-inflammatory hyperpigmentation.

The first reports of Sweet's syndrome (1964) highlighted its association with upper respiratory tract infections. Subsequent data suggested a relationship between the syndrome and myeloproliferative disorders, myelocytic leukemia, visceral malignancies, inflammatory bowel disease, connective tissue diseases, pregnancy, drug reactions, and other disorders (3). Because reported cases of NDDH are infrequent (fewer than 80 cases had been published by 2012), associations between this clinical entity and other diseases is less clear. Even so, conditions described as possibly being associated with NDDH are acute myelogenous leukemia, ulcerative colitis, metastatic lung cancer, B cell lymphoma, influenza vaccine, glomerulonephritis dialysis, streptococcus tonsillitis, chemotherapy, hypopharyngeal carcinoma, IgA gammopathy, Crohn disease, seropositive arthritis, sarcoidosis, hepatitis C, urinary

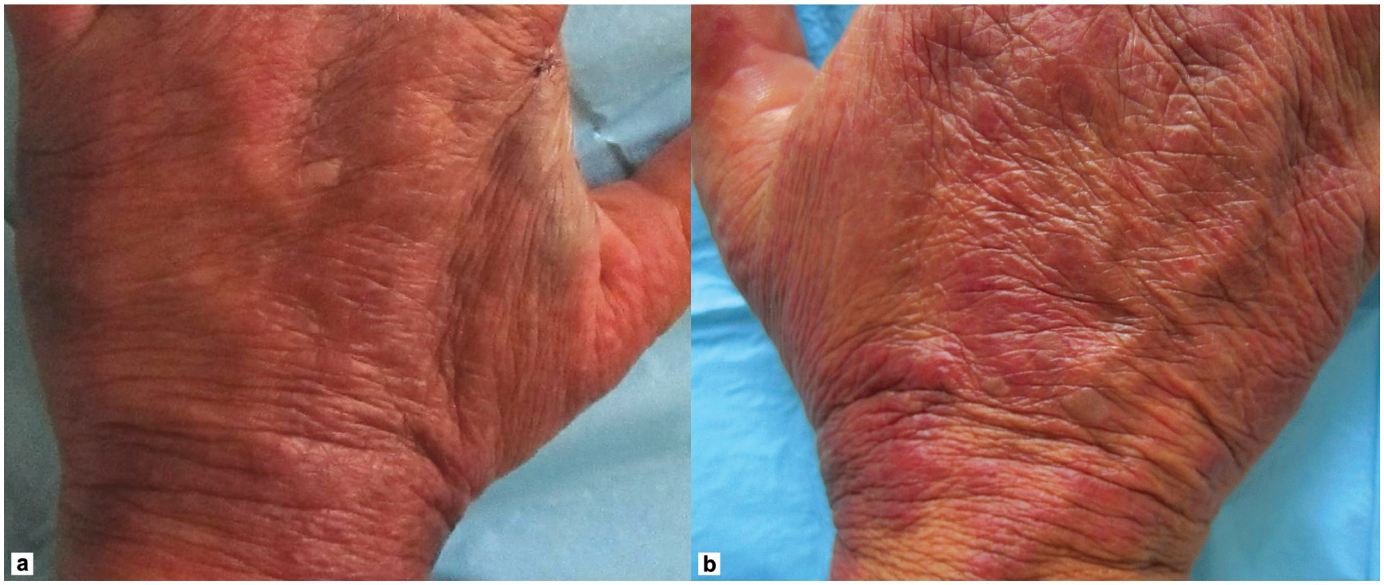

Figure 1 | a) Painful, erythematous, tumid, confluent papules and plaques, restricted to the dorsal left hand; b) Same aspects, right hand.

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tract infection, trauma, breast carcinoma, colon carcinoma, renal carcinoma, laryngeal carcinoma, thalidomide exposure, and fertilizer exposure (2). In our case, it is noteworthy that there was a previous diagnosis of polycythemia vera, which has not previously been reported with NDDH, but only described in association with the classic form of Sweet's syndrome (4).

Clinical presentation of NDDH differs from Sweet's syndrome in many aspects, in addition to the exclusive dorsal hand location. Fever, neutrophilic leukocytosis, and elevation of ESR have only been reported in one-third of patients, less significantly than what occurs in Sweet's syndrome (1). As in other series, other diseases were considered in the differential diagnosis, such as cellulitis or vesico-bullous pyoderma gangrenosum; this last entity has a recognized relation with polycythemia vera (5). However, the bilateral presence of skin lesions and its chronological evolution made those hypotheses unlikely and the histopathological aspects were highly suggestive of NDDH.

Currently, it is considered that the treatment of NDDH is the same as that of classic Sweet's syndrome, with many therapies having been suggested in the literature; namely: corticosteroids, dapsone, potassium iodide, colchicine, clofazimine, azathioprine, danazol, tetracyclines, and cyclosporine (3). Systemic corticosteroids are the most common first-line therapy. In our patient we decided to treat the condition only with a super-potent topical corticosteroid, which yielded a good outcome.

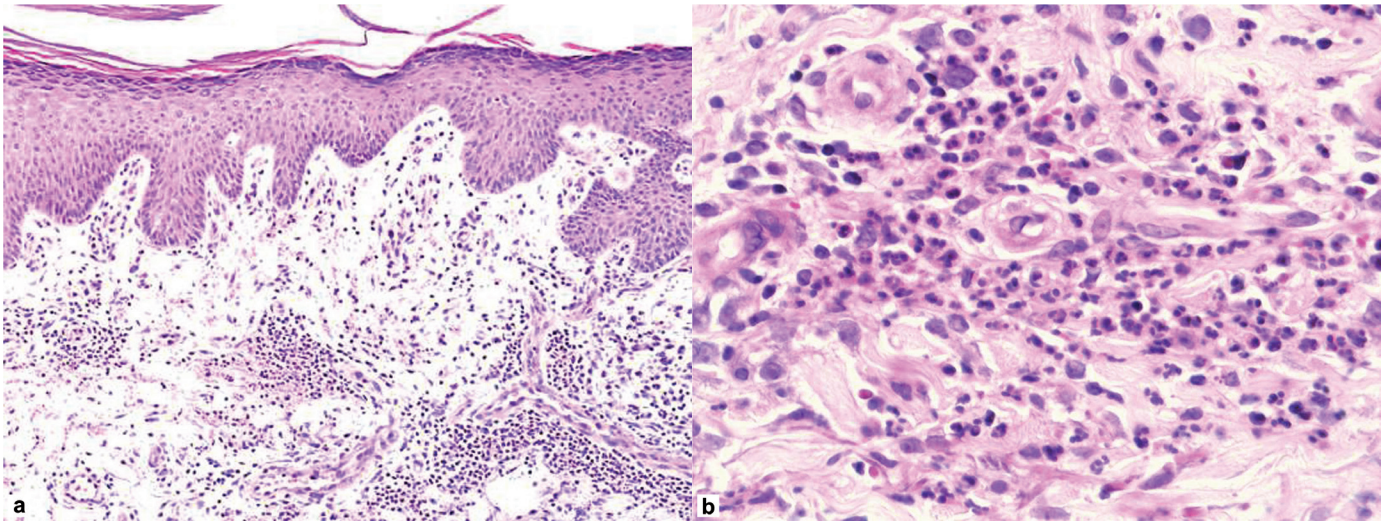

Figure 2 | a) Diffuse neutrophilic infiltrate on the papillary and middle dermis, predominantly perivascular, mainly composed of mature neutrophils (H&E, × 40); b) Perivascular neutrophil infiltration with some eosinophils, with no features of vasculitis (H&E, × 400).

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