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Images in Rheumatology

Case of idiopathic Sweet syndrome in young patient

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Sweet syndrome (SS) (acute febrile neutrophilic dermatosis) is a rare inflammatory syndrome characterized by the sharp appearance of painful, swollen, and erythematous papules, plaques, or nodules on the skin (1). SS is characterized by a sudden onset of fever, neutrophilia, and painful plaques. The classic histopathologic pattern of SS consists of a dense, diffuse neutrophilic infiltrate in the reticular dermis. In addition, there may be involvement of the eyes, internal organs, and the musculoskeletal system. Therefore, this disease is of significant interest to rheumatologists involved in the treatment of systemic pathology. SS is classified into three distinct categories: classical SS, malignancy-associated SS, and drug-induced SS (2). Classic SS most often develops after infections of the respiratory tract, inflammatory bowel disease, and pregnancy (3).

Here, we describe the idiopathic SS case of a young patient after acute respiratory infection. A 33-year-old patient complained of painful, dense rash on the skin of her shins, hips, back, trunk, and upper limbs, fever up to 38.6°C, myalgia, arthralgia, and general weakness. Initially, after a viral infection and hyperthermia, isolated rash appeared in the area of the knee joints; after 2 days, papules spread to the shins, hips, and back, and then, the temperature rose to 38°C. The patient had received antibacterial therapy, but there was not

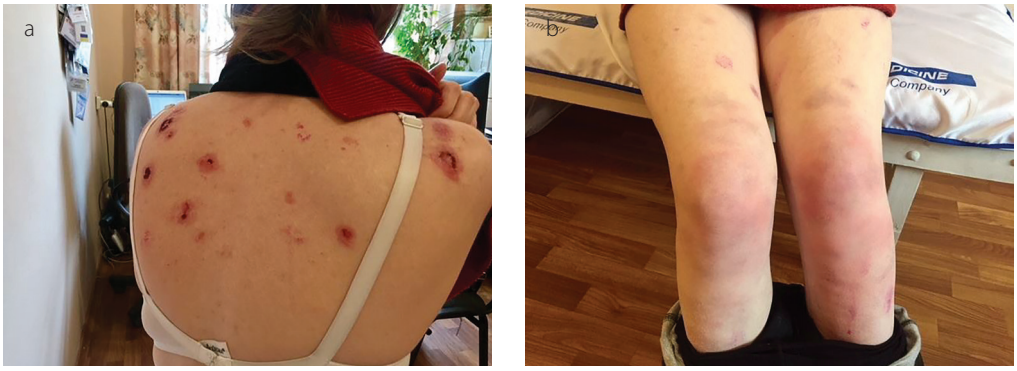

Figure 1. a, b. Dense erythematous nodules and papulose rashes on the back (a) and legs (b) with serous crusts and single vesicles, painless on palpation at the initial visit to the rheumatologist.

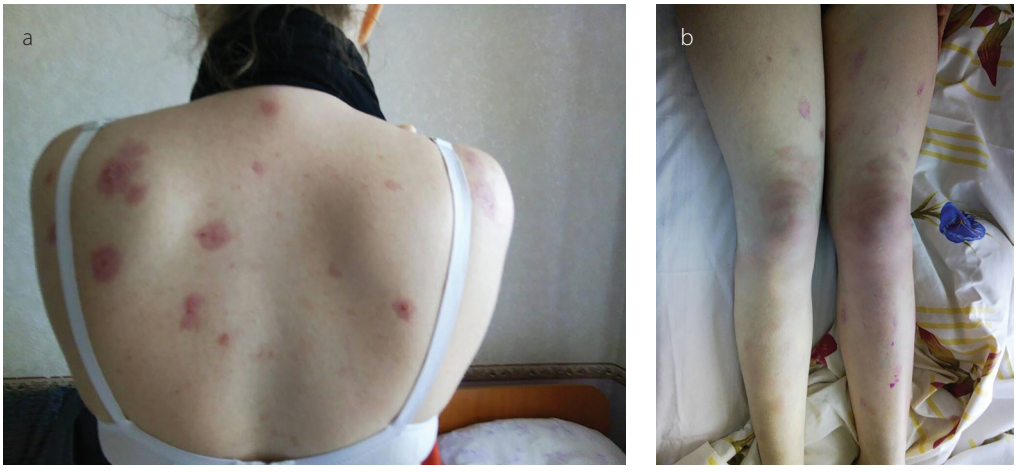

Figure 2. a, b. The dynamics of change in skin lesions one day after intravenous infusion of methylprednisolone. The decrease in the brightness and intensity of the rashes on the back (a) and legs (b) is clearly visible.

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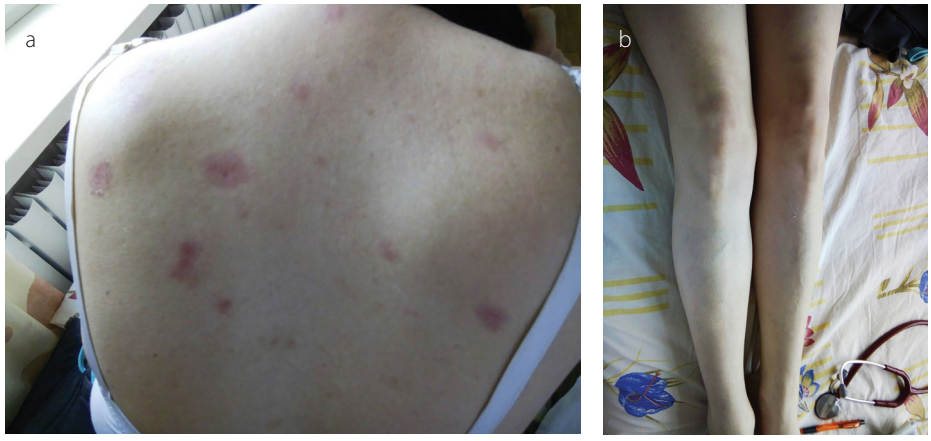

Figure 3. a, b. The dynamics of change in skin lesions on the back (a) and legs (b) after 2 months of therapy.

any benefit from this treatment. The skin rash continued to spread, and high fever was maintained. She consulted a dermatologist who suspected undifferentiated nodose vasculitis. The patient was referred to a rheumatologist.

An examination of the patient showed that there were multiple tender reddish blue or violet papules of various diameters (3-15 cm) in the shins and hips, confluent with indistinct boundaries. On the upper back and forearms, dense erythematous nodules, up to 3 cm in diameter, were present, as well as papulose rash with serous crusts and single vesicles, painless on palpation (Figure 1). The remainder of the examination was noncontributory. Laboratory examination revealed an increase in erythrocyte sedimentation rate (ESR) to 60 mm/h and C-reactive protein to 30 mg/L, anemia (hemoglobin 107 g/L), and neutrophilia (neutrophils 76%). Based on the rash characteristic, fever, increased inflammatory laboratory markers, and the exclusion of other diseases, a diagnosis of SS was made. Biopsy and histological examination were carried out, which confirmed our initial diagnosis of SS. According to the literature (4), a set of diagnostic criteria is used to diagnose SS. The major criteria are: 1) tender or painful erythematous plaques or nodules that appear suddenly, sometimes with vesicles, pustules, or blisters and 2) neutrophilic dermal infiltrate without signs of leukocytoclastic vasculitis. Minor criteria are: 1) nonspecific respiratory infection or infection of the gastrointestinal tract or vaccination; 2) fever >38°C; 3) the presence of three of the four laboratory deviations: white blood cell count >8000; ESR >20 mm/h; increase in C-reactive protein; and neutrophilia >70%; 4) good response to glucocorticoid treatment or potassium iodide. Our patient had two major and four minor criteria. SS can also be caused by inflammatory diseases such as chronic autoimmune disorders, solid malignant tumors, infections, hemoproliferative disorders, and pregnancy.

The patient was prescribed 125-mg intravenous methylprednisolone for 3 days with transition to oral administration -16 mg, followed by a gradual tapering off. One day after the start of glucocorticoid therapy, pronounced positive dynamics were noted (Figure 2). In 2 months, her condition resolved (Figure 3).

SS being a rare pathologic condition is of great interests to both rheumatologists and doctors of related specialties.

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