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Papular-purpuric "gloves and socks" syndrome in systemic mastocytosis treated with imatinib: a diagnostic challenge

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Mastocytosis is a rare disease characterized by the clonal proliferation of mast cells that accumulate in various body tissues and organs. Its classification comprises the cutaneous and systemic forms [1]. Imatinib is an oral tyrosine kinase inhibitor approved for the treatment of systemic mastocytosis (assuming the presence of KIT D816V mutation was excluded or its status is unknown).

A 47-year-old woman with indolent systemic mastocytosis (KIT D816V-negative) confirmed by bone marrow trephine biopsy (taken because she had multiple unexplained vertebral compression fractures) was admitted to the hospital due to erythematous lesions on the hands

and feet. Furthermore, the patient reported pain in the small joints of the hands and feet, as well as itching and tightness of the throat. She was treated with oral imatinib (600 mg per day) for 3 weeks due to the suspicion of clinically advanced mastocytosis (there was no other explanation for vertebral fractures at that time). The adverse drug reaction to imatinib was initially considered. The hematologist stopped imatinib and ordinated 20 mg of oral prednisone per day.

Because the skin lesions persisted for ten consecutive days, the patient was consulted with a dermatologist. Symmetrical erythematous, edematous plaques and peripheral papules were found on palms and the dorsal surfaces of hands and feet accompanied with swelling of the wrists and ankles. Additionally, there were follicular eruptions on the lower extremities, forearms, and elbows (Figure 1A-C). The dose of prednisone was increased to 40 mg per day for one week and additional laboratory tests were ordered.

The tryptase concentration was 18.00 µg/l (reference: 0.00-11.40 µg/l) and the activity of alkaline phosphatase was 127 U/l (reference: 35-104 U/l). Anti-nuclear antibodies were negative and the concentration of the C3c complement component was 0.86 g/l (reference: 0.90-1.80 g/l). Serological tests for HBV, CMV, and EBV were negative. The virus antibody panel showed positive results for parvovirus B19 with an elevated concentration of IgG (88.5 IU/ml, reference: <4 IU/ml) and IgM (2083 IU/ml, reference: <0.8 IU/ml).

At follow-up visits (after one and three weeks) the patient demonstrated flattening of the skin lesions, reduced swelling, and post-inflammatory exfoliation (Figure 1D-F).

Taking into account the clinical presentation and results of the auxiliary tests, the diagnosis of papular-purpuric “gloves and socks” syndrome (PPGSS) was made. The patient continued treatment of mastocytosis with imatinib at a previous dose.

PPGSS is an acute acral dermatosis that occurs primarily between the ages of 20 and 40. Its course is self-limiting and requires only symptomatic treatment [2]. This syndrome is

classically associated with parvovirus B19 infection [3]. The clinical presentation includes painful edema of the small joints of the hands and feet accompanied by pruritic erythematous papules with subsequent petechiae of "gloves and socks" distribution. Small ulcerations of the oral mucosa might coexist [3]. Laboratory test results are usually normal. In immunocompromised patients, PPGGS might be complicated by aplastic anemia, leukopenia [2], and thrombocytopenia [4]. Hypothetically, imatinib treatment could be a trigger for parvovirus B19 reactivation; however, a high titer of IgM antibodies suggests a primary infection that caused PPGSS.

This case highlights the need for a careful differential diagnosis of skin lesions initially suspected of being an adverse drug reaction to imatinib. Such eruptions might be observed in 10% to 90% of patients (depending on imatinib dose) [5].

Article information

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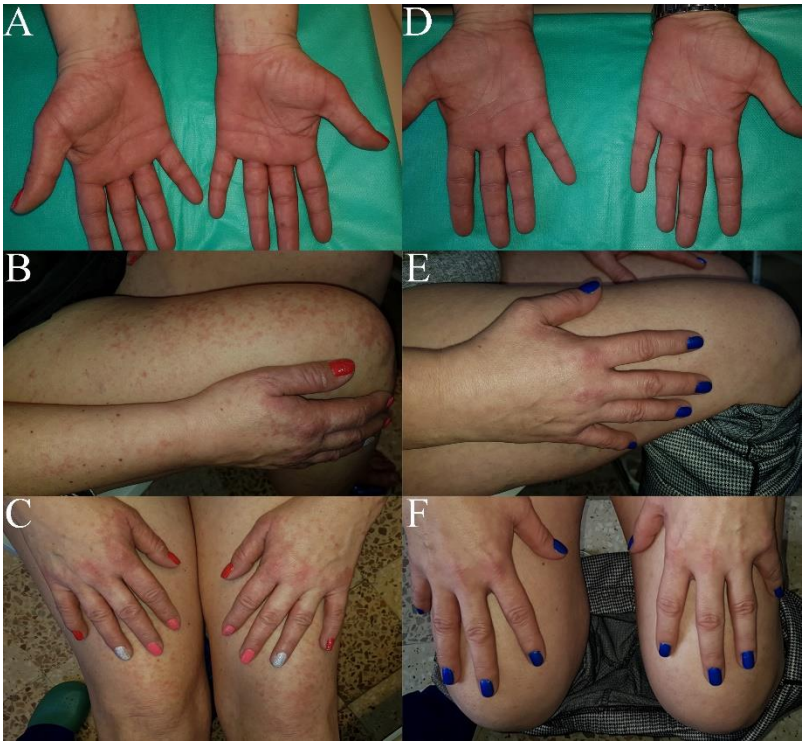

Figure 1 A-C – initial skin lesions: erythematous, edematous plaques and peripheral papules, localized symmetrically on palms and the dorsal surfaces of hands; follicular eruptions on lower extremities; D-F – visible improvement with reduction of edema and flattening of the skin lesions

Short title: Mastocytosis and popular-purpuric “gloves and socks” syndrome