A strange rash with "gloves and socks" distribution

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The papular-purpuric gloves and socks syndrome (PPGSS) is a new acral dermatosis first described in adults in 1990 and successively described in some children [1, 2]. Pruritic and painful edema and erythema are the characteristic features, with subsequent acral petechiae localized to the distal upper and lower extremities as gloves and socks distribution. Sometimes, it is associated with mucosal lesions and systemic symptoms such as fever, asthenia and lymphadenopathy [3].

A 4-year-old boy was admitted to the Department of Pediatrics of "SS Annunziata" Hospital, Chieti, Italy, due to fever lasting 2–3 days, associated with a pruritic erythematous rash localized predominantly to the upper and lower limbs.

On physical examination, the child showed erythematous and papular large itching lesions on the upper and lower limbs, characteristic of strophulus, and especially notable erythema and edema of the hands and feet, with a sharp demarcation at the level of the wrists and ankles of healthy skin (Figures 1 A and B). He did not show lesions of the oral mucosa, genital and conjunctival or hepato-splenomegaly.

Blood tests documented an increase of C-reactive protein (7.89 mg/dl) and erythrocyte sedimentation rate (35 mm/h), normal platelet (237 × 10^3/µl) and leukocyte count (7.47 × 10^3/µl), but with prevalence of peripheral blood mononuclear cells and eosinophils, associated with neutropenia. The other routine laboratory investigations, including electrolytes and hepatic and renal function, were normal. Allergy testing documented mild sensitization to cow's milk, egg white and grass pollen, with significantly increased values of total IgE (1483 kU/l). Serology (IgG and IgM) was negative for Epstein-Barr virus (EBV), cytomegalovirus (CMV), herpes virus (HSV) 1-2, rubella, Coxsackie virus, and toxoplasma. The quantitative real-time polymerase chain reaction (PCR) documented infection with parvovirus B19, with detection of IgM in the serum. Therefore, the diagnosis of PPGSS related to parvovirus B19 infection was made.

Papular-purpuric gloves and socks syndrome is an acute dermatitis that affects mainly young adults during the spring and summer. So far only a few pediatric cases have been reported [2, 4].

Characteristic of the disease at an early stage are pruritic erythema and edema, localized symmetrically on the hands and feet, with "gloves and socks" distribution, followed by an isolated or confluent erythematous papular and purpuric rash, with sharp demarcation at the wrists and ankles [4–6]. Similar lesions were observed in 50% of cases in the genital, oral, or other areas of the skin [1, 5], including petechiae, pharyngeal erythema, swollen lips, and painful oral erosions [6].

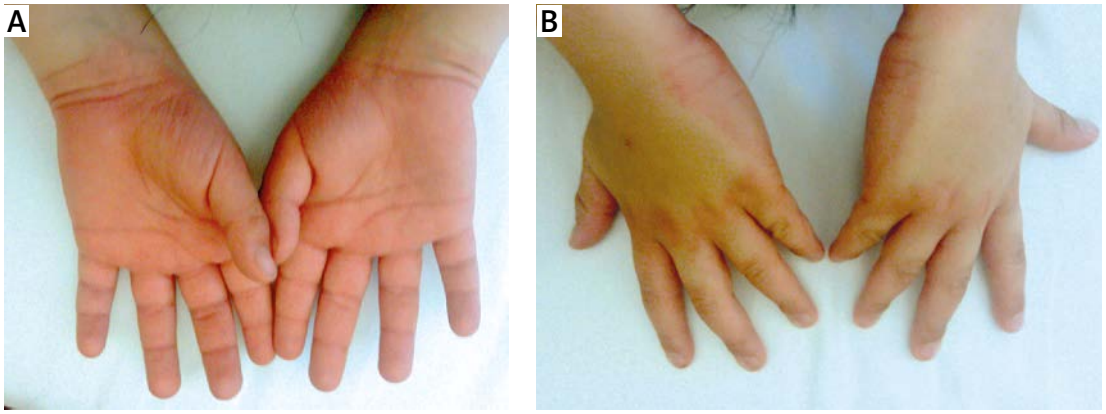

Figure 1 A, B. Erythema and edema of the hands with sharp demarcation at the level of the wrists of healthy skin

Gutermuth *et al.* could identify more than 70 cases documented in the literature; 66% of these were associated with parvovirus-B19 infection, transmitted by respiratory droplets. There was also sporadic association with other viruses such as varicella zoster, EBV, CMV, HSV 6-7, Coxsackie, hepatitis B, and rubella [5].

Prodromal or associated symptoms may be fever, asthenia, anorexia, arthralgia, myalgia, lymphadenopathy, lesions of the mucous membranes, and gastrointestinal symptoms [4, 6].

The symptoms are generally mild, especially in children and adolescents, while most severe cases occur in adults [5].

Resolution is usually spontaneous in 7–14 days, with slight peeling on palms and soles [4, 6].

The hypothesized pathogenesis is most likely due to immune complex deposition, supported by histopathology that usually shows lymphocytic perivascular infiltrates, edema of the papillary dermis, and extravasation of red blood cells; direct immunofluorescence indeed reveals IgM and C3 deposits at the dermo-epidermal junction in a granular pattern and in the walls of the papillary dermal vessels [4–8].

Serum IgM testing is recommended for diagnosis of acute infection in immunocompetent patients, demonstrating high sensitivity (89%) and specificity (99%) [9].

The symptomatic variety of PPGSS requires differential diagnosis with other rheumatologic and hematologic diseases: lymphoproliferative disorders, thrombocytopenic syndromes, Kawasaki disease, leukocytoclastic or hypersensitivity vasculitis (cryoglobulinemic vasculitis and Henoch-Schonlein purpura) and Still’s disease [9]. The many differential diagnoses also include Gianotti-Crosti syndrome, hand-foot-mouth disease, erythema multiforme and meningococcal sepsis [5].

Papular-purpuric gloves and socks syndrome is a rare manifestation of parvovirus B19 infection characterized by acute onset and a favorable, self-limited course. In most cases it needs only symptomatic treatment with antipyretics and non-steroidal anti-inflammatory drugs, avoiding the use of corticosteroids, because it is a self-limiting disease and immunosuppressive therapy may promote the persistence of the virus.

Since the first description in 1990, about 90 cases of PPGSS have been described in the literature, with 74 items reported in PubMed for papular-purpuric gloves and socks syndrome [9], while only 34 items result when adding the filter of age (birth to 18 years). There was only one report of this disease in Italian children [10], so to our knowledge this is the second report.

Papular-purpuric gloves and socks syndrome, although rare in children, must be taken into account as an unusual clinical manifestation of infection by parvovirus B19, which differs from the fifth disease (or infectious erythema), the much more common disease caused by this virus in children, mainly by location, characteristics of the rash and accompanying symptoms. A correct diagnosis is important to avoid unnecessary laboratory tests and treatment.

Conflict of interest

The authors declare no conflict of interest.

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