

## Papular-purpuric gloves-and-socks syndrome. Presentation of a clinical case

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### ABSTRACT

Papular-Purpuric Glove-and-Sock Syndrome is a rare, infectious disease, of viral etiology, characterized by the presence of pruritus, edema and symmetrical erythema, very well defined at the wrists and ankles with a gloves-and-socks distribution. Other areas can be affected, with a moderate erythema appearing in cheeks, elbows, knees, armpits, abdomen, groin, external genitalia, internal face of the thighs and the buttocks. Erosions, small ulcers, enanthema and blisters can be observed in the oral cavity and lips, and less frequently in other mucous membranes. Complications are rare, although they can be severe, 50% of the published cases are related with the Parvovirus B19. Due to its oral involvement stomatologists should be aware of this syndrome in order to carry out a correct diagnosis of the disease.

**Key words:** *Glove and Sock Syndrome, Parvovirus B19, purpura.*

### RESUMEN

El Síndrome Papular Purpúrico en Guante y Calcetín es un síndrome raro, infeccioso, de etiología, viral que se caracteriza por la presencia de prurito, edema y eritema simétricos, muy bien delimitadas a nivel de las muñecas y de los tobillos con el aspecto en guante y calcetín. Pueden afectarse otras áreas apareciendo un eritema moderado en mejillas, codos, rodillas, axilas, abdomen, ingles, genitales externos, cara interna de muslos y glúteos. En la cavidad oral y labios y menos frecuentemente en otras membranas mucosas se pueden observar erosiones, pequeñas úlceras, enantema y vesículas. Las complicaciones son raras aunque pueden ser severas El 50% de los casos publicados están relacionados con el Parvovirus B19. La afectación oral en este síndrome hace que deba ser conocido por los estomatólogos para realizar un correcto diagnóstico de esta enfermedad.

**Palabras clave:** *Síndrome en Guante y Calcetín, Parvovirus B19, púrpura.*

## INTRODUCTION

Papular-Purpuric Glove-and-Sock Syndrome is a rare disease, usually of infectious etiology, which, in addition to a cutaneous involvement, manifests with lesions in the oral cavity.

In 1990 Harms, Feldman and Saurat described, in 5 Swiss patients (1), a papular dermatosis characterized by edema and erythema that evolved toward a petechial purpura with a characteristic glove and sock distribution. The etiology was unknown, although the authors suggested an infectious origin.

In 1991, Bagot and Revuz were the first authors to implicate the Parvovirus B19 in the etiology of this syndrome (2), and since then numerous studies have indicated its involvement in more than 50% of cases (3,4,5,6,7).

Parvovirus B19 is the only parvovirus that produces pathology in humans; it is a DNA virus of the Parvoviridae family that infects the precursor cells of erythrocytes in bone marrow. It is usually contracted via respiratory secretions, fundamentally in childhood, and frequently produces a type of febrile exanthem known as erythema infectiosum or fifth disease (8). It can also produce acute or chronic arthropathies, fetal hydrops, abortions, aplastic anemias (9), hepatitis (10) or, much less frequently, Sjögren's syndrome (11) or papular-purpuric glove-and-sock syndrome.

This syndrome affects children or young adults, appearing equally in both sexes and appears fundamentally in spring and summer. The rash is usually accompanied by fever and is self limiting, disappearing in one or two weeks with a slight desquamation of the areas involved. Only symptomatic treatment is required, relapses having not been described.

In the oral mucosa it presents as multiple petechiae on the hard and soft palate and as small erosions in the oral mucosa and tongue; commissural cheilitis can also present.

Nonspecific urethritis can appear as other mucosal involvement.

Diagnosis of this syndrome is made by the clinical dermatological characteristics and is confirmed by specific serology for Parvovirus B19 using enzyme-linked immunosorbent assay (ELISA), or polymerase chain reaction (PCR). Active infection is demonstrated by high IgM titers that subside over 2-4 weeks when an elevation in IgG appears.

The differential diagnosis should be made against urticaria-angioedema, serpiginous purpuric eruption of hands and feet, hand-foot-and-mouth disease, exanthem subitum, or erythema infectiosum, which are also produced by Parvovirus B19 (12).

## CLINICAL CASE

A 9 year-old male referred by his pediatrician for presenting a possible aphthous stomatitis accompanied by pruriginous lesions on the feet, hands and buttocks of 48 hours evolution.

On examination the patient appeared to be in generally very good health, although with febricula, and with petechial lesions in the oral cavity and erythema on the palate (Figure 1) as well as aphthous lesions appearing on the floor

of the mouth and ventral face of the tongue (Figure 2), and the same type of lesions on the soft palate (Figure 3). The dermatological examination revealed a multitude of papular lesions of purpuric appearance on the feet (Figure 4) and buttocks, these same lesions appearing much less numerous on the hands and axillae.

Likewise, the patient presented a urethritis with a slight purulent exudate from which a culture was made, resulting as normal flora.

Analytical tests were made, finding only a moderate platelet deficiency as well as serology to Herpes virus type I and II; Coxsackievirus and Cytomegalovirus, which were negative, and Parvovirus B19 being in this case IgM positive and IgG negative. Treatment with paracetamol was established.

At 10 days the patient was completely reestablished, all lesions, both oral and cutaneous, having disappeared without sequela.

For reasons beyond our control (summer vacations, beginning of school etc.) several months passed before seeing the patient again, at that time taking the opportunity to repeat the serology to Parvovirus, the IgG titers being very high while those of IgM had substantially subsided.

In view of the clinical picture and of the patient's seroconversion for Parvovirus B19, a diagnosis of Papular-Purpuric Glove-and-Sock Syndrome was established.



Fig. 1. Numerous petechiae on the palate.



Fig. 2. Small erosions and erythema on the ventral mucosa of the tongue.



Fig. 3. Lesions on the soft palate.



Fig. 4. Purpuric papules with sock distribution.

## DISCUSSION

Papular-Purpuric Glove-and-Sock Syndrome is a rare clinical form of infection by Parvovirus B19. Since its description in 1990 by Harms, and since Bagot related it with this virus in 1991, this etiology has been demonstrated in 50% of the described cases. In other cases, other virus such as Herpes Virus type 6 (13) Cytomegalovirus (14,15) or Coxsackie B6 (13), bacteria such as *Arcanobacterium haemolyticum* (16) or drugs such as trimethoprim-sulfamethoxazole (17) have been implicated.

The pathogeny of this illness is not perfectly clarified, but in the case of Parvovirus infection, viral particles have been isolated in vessel endothelium and in basal cells of the epidermis at the time of the exanthema. In spite of this, it is debated as to whether the clinical manifestations are a consequence of the viremia (18) or of circulating immunocomplexes (19).

The patient presented here constitutes a characteristic case of this syndrome, for reason of its typical clinical manifestation, its appearance in spring and a perfectly demonstrated seroconversion.

It is important to carry out a differential diagnosis against hematological disorders, such as aplastic anemia or Henoch-Schönlein purpura whose clinical and analytical findings will be conclusive. Likewise, against viral infections such as measles, which affect the whole body and do not have the characteristic distribution of this syndrome, hand-foot-and-mouth disease, and infectious mononucleosis above all if accompanied by cutaneous rash or herpangina. Some

dermatologic syndromes such as Gianotti-Crosti Syndrome or Kawasaki disease should be included in the differential diagnosis.

In conclusion, Papular-Purpuric Glove-and-Sock Syndrome is a self-limiting acrodermatosis, of unknown pathogeny that fundamentally affects children and young adults. It is characterized by edema and pruritus of hands and feet, followed by a purpura at the same site, involvement of the oral mucosa with erythema and petechiae, and some minimal alterations in the analyses. Parvovirus B19 is implicated as the etiologic agent in 50% of the described cases, but other etiologic agents can also be involved.

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