Perifollicular Palpable Purpura as the Initial Manifestation of HIV Infection

To the Editor:

Leukocytoclastic vasculitis is a disease associated with many causes.1 It presents with palpable purpuric lesions on the lower limbs. In patients with human immunodeficiency virus (HIV) infection it has been reported that these lesions tend to have a perifollicular localization.2-4 Case 1. A 28-year-old homosexual man with no relevant history presented to the emergency department with intermittent pruritic lesions on the legs that had started 2 months earlier and were associated with arthralgia in the knees and ankles. Physical examination showed predominantly perifollicular purpuric papules on the anterior aspect of both legs (Figure 1). No symptoms suggestive of scurvy, such as gingival bleeding, hyperkeratotic papules, or hair changes, were observed. A skin biopsy showed leukocytoclastic vasculitis (Figure 2). Additional tests detected positive serology for HIV type 1. The total CD4 count was 120/µL with a CD4:CD8 ratio of 0.13. Immunoglobulin (Ig) G and IgA levels were increased, with values of 2120 mg/dL and 1130 mg/dL, respectively. Other additional tests (chest radiograph, Mantoux, complete blood count, biochemistry, urinalysis, complement levels, vitamin C levels, autoantibodies, rheumatoid factor, IgM, IgE, cold agglutinins, and serology for syphilis, hepatitis B and C virus, cytomegalovirus, toxoplasma, herpes simplex virus, and varicella-zoster virus) were normal.

Figure 1 Perifollicular purpuric lesions on the legs in case 1.

References

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started 3 days earlier. Five days earlier he had started treatment with amoxicillin-clavulanic acid for recurrent throat infection. He denied having other symptoms in the systems review. Physical examination showed perifollicular purpuric papules on the back of both feet and legs. A skin biopsy revealed leukocytoclastic vasculitis and direct immunofluorescence was weakly positive for IgA deposits in some superficial vessels. HIV serology was positive for HIV-1. The CD4 count was 400/µL with a CD4:CD8 ratio of 0.45. IgG and IgA levels were increased, with values of 210 and 310 mg/dL, respectively. The remaining additional tests were normal.

Leukocytoclastic vasculitis is uncommon in HIV infection and, when it does occur, the triggering factors are usually the same as those found in the general population (infection, systemic disease, drugs, etc.). In a number of patients no causal agent is found and the vasculitis is considered to be a direct result of the virus infection. In these cases, 3 specific mechanisms have been proposed: 1) immune complex deposition facilitated by increased serum immunoglobulin levels, 2) T cell-mediated damage to the blood vessels secondary to oligoclonal CD8 expansion, and 3) direct infection and replication of the virus itself in the vessel wall and in the perivascular region.

Although the clinical presentation of leukocytoclastic vasculitis is fairly homogeneous, it has been observed that palpable purpura tends to have a perifollicular localization in HIV-infected patients. Barlow et al described a 20-year-old homosexual man whose first manifestation of HIV infection was generalized lymphadenopathy and follicular purpuric papules on the trunk and extremities. Two subsequent reports showed that HIV infection can cause perifollicular purpura in the absence of other triggering factors and that it may also favor follicular accentuation of vasculitis of a different origin.

Although several hypotheses have been proposed, the cause of the follicular localization of purpura in HIV is unknown. It has been suggested that the virus itself could cause vitamin C depletion. Another hypothesis is that subclinical vascular damage in the area of the follicle affects immune complex deposition at this level.

Scurvy should be included in the differential diagnosis of perifollicular purpuric lesions. Perifollicular hemorrhage is one of the earliest and most specific signs of this nutritional deficiency. Obtaining other clinical data, determination of vitamin C levels, and dermoscopy can help to differentiate them.

In conclusion, HIV infection should be included in the differential diagnosis of perifollicular palpable purpura. We report 2 patients whose first manifestation of the infection was perifollicular leukocytoclastic vasculitis. This manifestation led to a clinical suspicion of infection, which was subsequently confirmed by serology.

References
Pachydermodactyly: A Rare Form of Acquired Digital Fibromatosis

Paquidermodactilia: una forma poco frecuente de fibromatosis digital adquirida

To the Editor:

Pachydermodactyly is a rare, benign form of acquired digital fibromatosis, characterized by soft tissue swelling that affects the skin of the fingers, specifically on the lateral aspect of the proximal interphalangeal (PIP) joints, mainly of the second, third, and fourth fingers. It particularly affects adolescents with no family history and is occasionally confused with rheumatological diseases.

To date, few cases have been reported in the medical literature; we describe a new case.

The patient was a 14-year-old boy with no past history of interest who was referred to the dermatology clinic for evaluation of diffuse thickening that had developed some months earlier on the second, third, and fourth fingers of each hand. A point of interest is that the patient was a competition-level climber who practiced for several hours each day.

Physical examination revealed bilateral and symmetric swelling of the lateral aspect of the PIP joints of the second, third, and fourth fingers of each hand (Figures 1 and 2), more marked on the right side (Figure 2). There were no other relevant alterations of the skin.

A skin biopsy of one of the affected areas revealed a thickened dermis with an increase in the number of collagen fibers and a slight increase in the number of fibroblasts. There was also a slight increase in the quantity of mucin, with no significant inflammatory infiltrate. The overlying epidermis was hyperkeratotic with compact orthokeratosis and thickening of the dermis (Figure 3).

Blood tests were requested, including kidney, liver, and thyroid function, rheumatoid factor, antinuclear antibodies, and other autoimmune studies, all with normal or negative results.

Plain radiography of the hands revealed thickening of the soft tissues around the PIP joints, with no bone or joint abnormalities (Figure 4).

Pachydermodactyly was first described by Bazex in 1973 and it was named by Verbov 2 years later. It is a rare disease, though its true incidence could be underestimated. This benign fibromatosis mainly affects adolescent men and the diagnosis is questionable if made in adults. In many cases there is a history of manual sporting practice such as martial arts, weight-lifting, or climbing (as in our case).

The etiology is unknown. Rai and Zaphiropoulos suggested that it might represent an incomplete presentation of pachydermoperiostosis. Pachydermoperiostosis or Touraine-Solente-Golé syndrome is the primary or idiopathic form of hypertrophic osteoarthropathy. The secondary form is more common and usually follows lung or heart disease, which may be of a neoplastic nature. Pachydermoperiostosis is defined by 3 major criteria: pachyderma, periostosis, and clubbing of the fingers. There are also minor criteria (seborrhea, sebaceous hyperplasia, folliculitis, acne, and more). The primary form is considered to be a hereditary disease, although a family history is found in only 25% to 38% of cases. In addition, it has a variable degree of penetration, and complete forms of the syndrome are therefore uncommon.

Clinically, pachydermodactyly is characterized by asymptomatic symmetric swelling of the PIP joints, predominantly on their lateral aspect, and particularly affecting the second, third, and fourth fingers. Occasionally it also affects the dorsum of the hand, then receiving the name pachydermodactyly transgrediens.

Histopathology study reveals epidermal hyperplasia with compact orthokeratosis and thickening of the dermis with an increase in the number of collagen fibers and a mild proliferation of fibroblasts, with no significant inflammatory infiltrate.

The differential diagnosis should include certain forms of polyarthritis that can affect the PIP joints, such as the polyarticular form of juvenile chronic arthritis, psoriatic arthritis, and rheumatoid arthritis. However, all those disorders have characteristic joint involvement, whereas pachydermodactyly is an asymptomatic fibrous swelling of the PIP joints of the second, third, and fourth fingers with no bone or joint abnormalities.

Pachydermodactyly must also be differentiated from juvenile hyaline fibromatosis, although there are some authors who, as occurs with pachydermoperiostosis, consider that pachydermodactyly is a localized form of juvenile hyaline fibromatosis. Juvenile hyaline fibromatosis is a mesenchymal dysplasia of autosomal recessive inheritance that appears in early childhood or in adolescence; only about 65 cases have been reported. It is characterized clinically by skin lesions, gingival hypertrophy, flexion contractures of the large joints, and bone lesions. The skin lesions consist of multiple tumors, often located on the scalp and around the nose, associated with small pearly papules and plaques on the trunk, chin, and ears and around the nares. Juvenile hyaline fibromatosis is due to synthesis of abnormal collagen, which is deposited as