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ERYTHEMA ELEVATUM DIUTINUM, A CHRONIC CUTANEOUS VASCULITIS, IS ASSOCIATED WITH MONOCLONAL GAMMOPATHY AND DETERMINES CHRONIC RHEUMATOID-LIKE POLYARTHRITIS

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BACKGROUND

Erythema elevatum diutinum (EED) is a chronic and rare form of leukocytoclastic vasculitis, manifesting with papules or nodules of violet to brownish-red coloration, with predominance in extensor surfaces of joints. The etiology remains unknown. The EED has been associated with rheumatoid-like polyarthritis and with gammopathy monoclonal IgA. Histopathological study of skin lesions is essential for the diagnosis.

CASE REPORT

Male patient, 45 years old, chronic smoker, reports that seven years earlier he started having pain in multiple joints. The articular course has become chronic, determining progressive deformities in hands and wrists, with swan neck deformity (Figs. 1 and 2). In the past three years, he had begun with a skin condition that worsened with lesions in plaques and erythematous-violet nodules, in the extensor face of the forearms and elbows (Fig. 3). He also developed purpuric lesions as petechiae in his legs two years earlier (Fig. 4). The skin biopsy confirmed vasculitis urticaria. The exams showed absence of antibodies, such as rheumatoid factor and anti-CCP; as well other antibodies such as ANF, anti-SSA, anti SSB, anti-DNA, anti-SM and anti-RNP were also negative. Skin biopsy of nodular lesions of the right forearm reviews histological changes compatible with leukocytoclastic vasculitis. Also, in the laboratory exams, covering complement components (within the normal range), IgA of 375 (VN < 350), IgE of 17, IgA lambda monoclonal immunofixation, IgG 864, IgM 30 (VN 50–300), light chain research free lambda 134 (VN 5.71–26.3), KAPPA 11.27 (VN 3.3–19.4) and K/L ratio 0.08 (VN 0.26–1.65). Although showing visible joint deformities, the X-ray showed an absence of erosion in the joints, with extensive deformities. The patient tried some treatments that didn't work as expected, like prednisone, methotrexate, leflunomide, adalimumab, golimumab and tocilizumab, with no improvement in the joint condition and persistence of the cutaneous condition. This case is an unusual presentation of a rare disease with serious debilitating consequences.

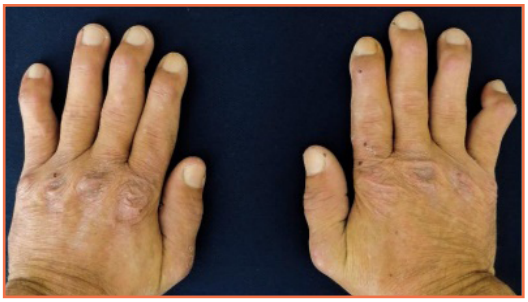

Figure 1. Swan neck deformity in hands.

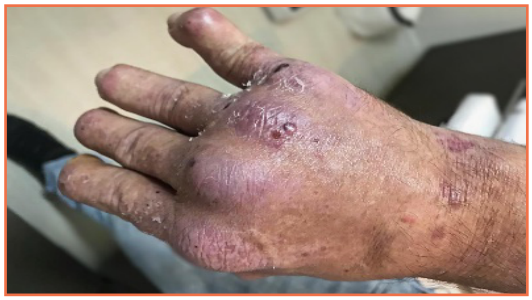

Figure 2. Swan neck deformity in hands.

Figure 3. Erythematous-violet elbow lesion.

Figure 4. Purpuric lesion as petechiae in leg.

CONCLUSION

Erythema elevatum diutinum should be considered in patients with complaints and/or rheumatological signs, such as long-standing arthralgias and increased evidence of inflammatory activity, associated with characteristic skin manifestations and a compatible histopathological study. Our case report confirms the association between EED and IgA monoclonal gammopathy and informs us that joint involvement can suggest rheumatoid arthritis.