Rare presentation of erythema elevatum diutinum

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INTRODUCTION

Erythema elevatum diutinum (EED) is a rare, chronic, and progressive skin condition, classified within the neutrophilic vasculitides. It presents as erythematous and violaceus papules and plaques. The distribution tends to be bilateral and symmetrical, overlying joints and localized on extensor surfaces. EED generally affects middle-age patients. Although its cause is yet unknown, it can be associated with other underlying autoimmune, infectious, or malignant diseases. Dapsone is used as a first-line therapeutic agent. We present a rare case of EED in terms of its severity and the high functional impact it caused.

CASE REPORT

An 83-year-old man presented with voluminous and extensive painless nodules on his limbs. He had neglected the lesions for more than 3 years, and they gradually spread and grew, making prehension and everyday gestures strenuous. The patient was otherwise fit and well. Physical examination found raised, well-defined, deforming violaceus hard nodules. These were distributed on the palms, on both anterior and posterior sides of the fingers, and on the elbows, knees, and soles (Fig 1). Results of laboratory tests including full blood count, renal function, and liver profile were within normal range. HIV, hepatitis B and C serology were negative. Electrophoresis of serum proteins showed the presence of a monoclonal IgG λ (8 g/L; normal limits, 0.4-4.2 mg/L) (Fig 2). A punch biopsy for hematoxylin-eosin was performed and showed a dermal nodule made of a dense neutrophilic infiltrate admixed with large collagen bundles and neoangiogenesis.

Abbreviation used:

EED: erythema elevatum diutinum

Together with the distribution and clinical features, histology supported the diagnosis of EED (Fig 2). To start dapsone, the patient underwent a glucose-6-phosphate dehydrogenase screening, which found an enzymatic deficiency; therefore, the drug was contraindicated. He was then lost to follow-up.

DISCUSSION

EED classically spares palms and soles, and the atypical tumoral appearance led us to consider, initially, differential diagnoses such as Kaposi sarcoma, secondary metastases of a malignant process, or a granulomatous reaction such as rheumatoid nodules or cutaneous sarcoid. First described in 1878 by Hutchinson, EED's physiopathology is still not clearly defined. It is thought to be a small vessel vasculopathy triggered by immune complexes. Although idiopathic forms have been described, it is usually associated with a wide range of underlying systemic conditions: autoimmune diseases (such as diabetes, rheumatoid arthritis and coeliac disease), chronic infections and malignancies (multiple myeloma, hematologic malignancies, or breast cancer). Drug-induced EED has also been described in rare cases. To date, we have found only 8 reported cases of palmoplantar EED, indicating that it is rather rare. Four of the 8 cases were associated with monoclonal gammopathy of undetermined significance[1-3] or multiple myeloma.[4] Two cases were not

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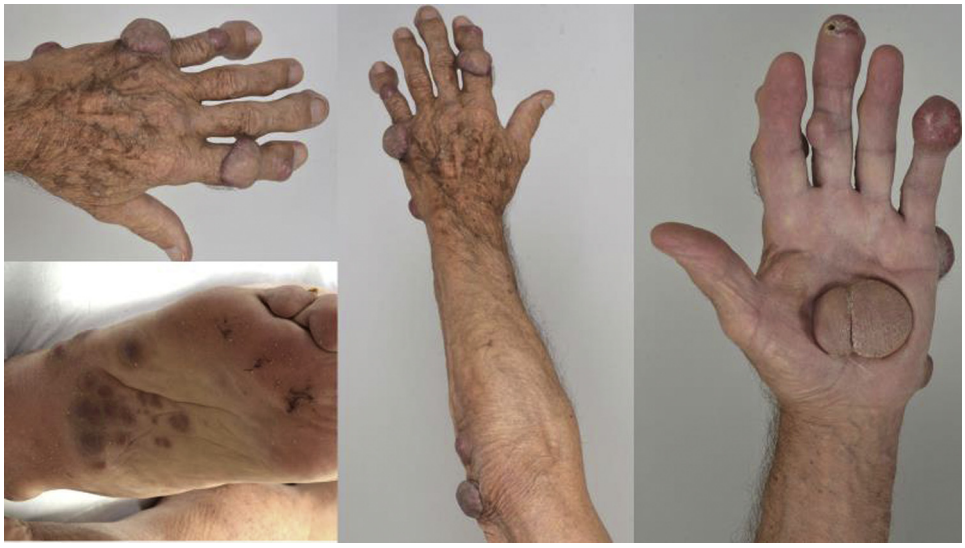

Fig 1. Clinical presentation. Extensive tumorlike nodules distributed on the dorsum of the hands, palms, soles, and elbow.

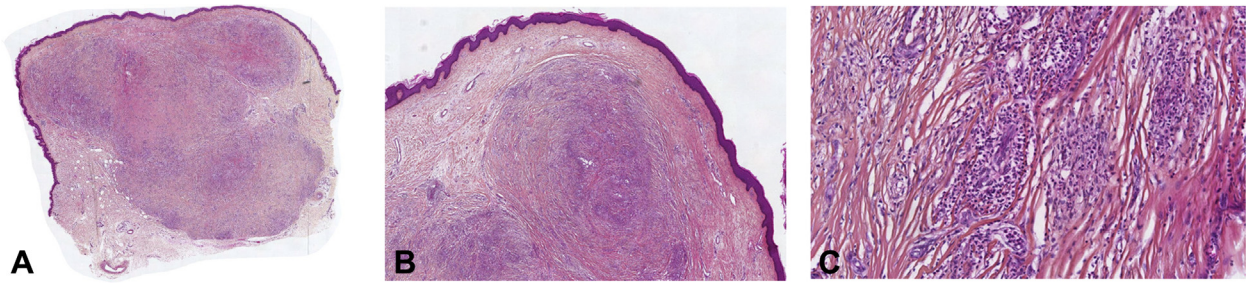

Fig 2. The sections show a raised dermal nodule made of a dense neutrophilic infiltrate admixed with large fibrosing collagen bundles in a storiform pattern and neoangiogenesis. The epidermis as well as the adnexal structures appear clear. (Original magnifications: **A**, ×2.5; **B** ×10; **C** ×20.)

concomitant with an underlying disease, 1 was found associated with Behcet disease,[5] and 1 was associated with HIV and hepatitis C and B viral co-infections.[6] In the reported palmoplantar cases, ages ranged from 32 (HIV patient) to 77, with an average of 54 years. We are not able to explain why, in our case, the patient, at 83 years of age, was considerably older than the other patients studied.

Screening for monoclonal gammopathy is recommended for all patients with EED,[7] as paraproteinemia is strongly associated with the condition.[8]

Although we planned to measure the serum-free light chains, our patient was lost to follow-up and we were not able to carry out the investigation. Taking into account his age and the fact he did not present with anemia or bone pain, no other investigation was conducted.

Several therapeutic options are described in the literature, including dapsone, tetracyclines, colchicine, nicotinamide, cyclophosphamide, and corticosteroid injections.[9] Dapsone tends to offer the best results but was contraindicated in our case because of the glucose-6-phosphate dehydrogenase deficiency. Surgical management with regular interventions is also possible in severe and advanced cases. Management of the underlying disease is usually attempted in EED patients. Asymptomatic monoclonal gammopathy of undetermined significance does not always require specific treatment. However, a 2014 review article revealed that treating the underlying gammopathy often showed clinical benefits for the skin[9] and should be considered as a therapeutic option. In our case, treating the monoclonal gammopathy seemed too aggressive for this elderly patient.

This case is an unusual presentation of a rare disease with serious debilitating consequences.

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