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Case Report

Erythema elevatum diutinum: a rare case of atypical presentation involving palms

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ABSTRACT

Erythema elevatum diutinum (EED) is a rare chronic dermatosis which mostly affects adult individuals. The classical skin lesions are asymptomatic, plum colored, erythematous nodules and plaques of variable sizes distributed symmetrically over extensor aspect of extremities. We reported a case of EED in a 50-year-old female presenting with lesions involving palms owing to its rare presentation.

Keywords: EED, Chronic dermatosis, Asymptomatic, Plum-colored nodules, Extensor aspect of extremities

INTRODUCTION

The majority of patients with Erythema elevatum diutinum (EED), a rare chronic dermatosis, are adults. It is distinguished by red-violet to red-brown papules, plaques, and nodules that prefer the extensor surfaces.[1,2] Hutchinson and Bury first identified the disorder in the 1880s, and Radcliffe Crocker and Williams gave it a name in 1894.[3] Asymptomatic (rarely painful), plum-colored erythematous nodules, and plaques of various sizes are the distinctive skin lesions. They are distributed symmetrically over the extensors of the extremities, particularly over the joints, the dorsal aspect of the hands, feet, and elbows, the buttocks, and the Achilles tendon, as well as the buttocks and face occasionally.[2] Here, we described the case of a 50-year-old woman who presented with an unusual form of EED affecting both palms.

CASE REPORT

A 50-year-old female presented to the out patient department with asymptomatic lesions over both palms, dorsum of hands and feet since 1 month. The lesions were in the form of multiple, well-ill defined, dusky plum colored erythematous and violaceous to hyperpigmented, macules, papules with a few nodules. They were progressive in size and some of them showed resolution with hyperpigmentation. There was no deep dermal tenderness associated with the lesions over palms. She had no associated arthralgia or arthritis. She did not experience any constitutional symptoms, eye complaints or sore throat. She was advised some investigations including blood cell counts, urine analysis and venereal disease research laboratory (VDRL) titers which were found to be within normal limits. Based on the history and clinical examination a provisional diagnosis of EED with a differential diagnosis of atypical erythema multiforme was made and a skin biopsy was advised to confirm the diagnosis. Epidermis showed mild hyperkeratosis with wavy lower border and pigmented basal layer. Dermis showed perivascular lymphocytic infiltrate. Few dilated blood vessels were seen and neutrophils were seen infiltrating the wall at places. Dermal oedema was also seen. The above findings confirmed the diagnosis of EED. As the lesions were mild and asymptomatic the patient was prescribed oral anti histaminic and topical emollients to which she responded well.

Figure 1: Multiple plum coloured to hyperpigmented macules over bilateral palms.

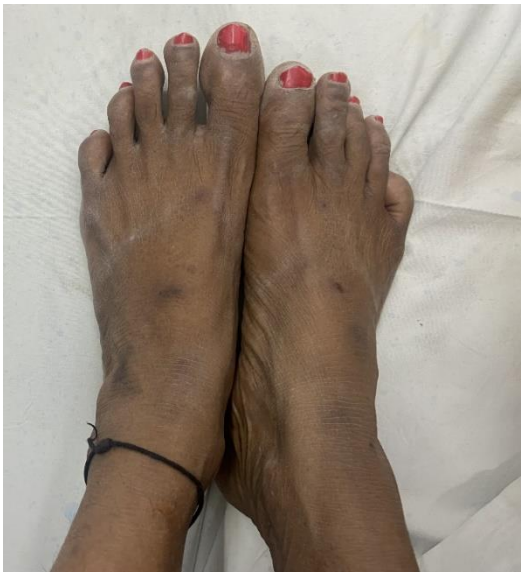

Figure 2: Multiple plum coloured to hyperpigmented macules, papules and few nodules over dorsum of bilateral feet.

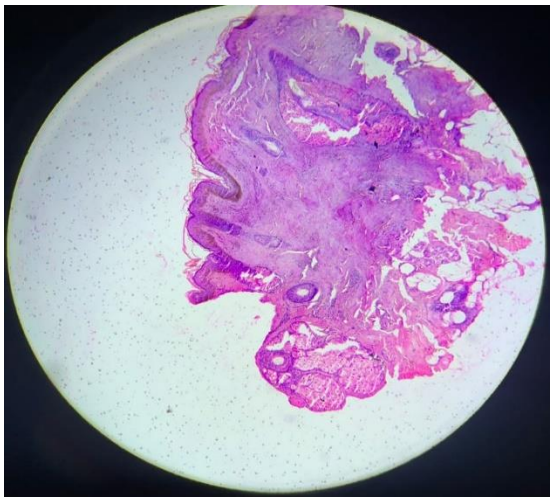

Figure 3: Histopathology of EED.

DISCUSSION

Hutchinson et al discovered EED first in 1888 and 1889, respectively. Radcliff-Crocker et al first used the term EED after noticing similarities between their cases and those of Bury et al.[4] Asymptomatic, plum-colored to erythematous papules, nodules, and plaques of various sizes are distributed symmetrically over the extensors of the extremities, the dorsum of the hands and feet, the buttocks, elbows, knees, the Achilles tendon, and occasionally over the face and ears.[2] Patients may occasionally feel discomfort or a burning feeling over the lesions. The patient may experience arthralgia, fever, or other extracutaneous signs. Despite being benign, the condition may also be linked to autoimmune illnesses, infections, or hematological abnormalities, among other things. By 5-10 years, the disease might spontaneously improve.[1]

EED can appear at any age, but it was most frequently diagnosed in people in their fourth to sixth decades, with no racial or gender variations. The start may occur sooner in an HIV-positive person than in a healthy person. Although the exact origin of EED is unknown, it is believed to develop as a result of immune complex deposition in dermal blood vessels, which causes inflammation as a result of complement fixation. The epidermis research demonstrates the deposition of immune complexes in small vessels, which activates complement, attracts neutrophils, and emits corrosive enzymes. It's possible that antineutrophilic cytoplasmic antibodies cause EED. The beginning of EED is thought to involve the activation of cytokines like interleukin-8, which causes the selective recruitment of leukocytes to blood vessels, resulting in repeated injury to the vessels and fibrosis. In the later phases of the illness, this ultimately leads to the deposition of fibrin in and around small dermal vessels.[1] Two out of every five patients in Katz et al research demonstrated the prevalence of recurrent bacterial infections brought on by *Streptococcus* spp. Three out of five patients also demonstrated increased C1q binding, indicating the existence of circulating immune complexes.[1,5]

The upper and middle dermis of EED exhibit fibrin deposition and early leukocytoclastic vasculitis changes with polymorphonuclear cell infiltrate. The blood arteries are surrounded by a leukocytic infiltrate, macrophages, and histiocytes. Along with capillary oedema, these infiltrates may build up between collagen bundles and manifest clinically as pseudovesiculation. Spindle cell growth and granulation tissue can both be seen. In the fibrotic tissue, extracellular cholesterol crystals may be seen. Direct immunofluorescence may show vasculitic alterations such as immunoglobulins, complement, and intra- and perivascular fibrin deposition (Ig G, IgA, IgM).[1]

The extensor surfaces of the limbs are covered in asymptomatic, symmetrically dispersed erythematous to

violaceous papules and nodules that are solitary or confluent with a hardened consistency. However, reports of typical lesions at atypical locations and vice versa.[6] Rare instances can result in constitutional symptoms, arthralgias, scleritis, uveitis, autoimmune keratolysis, and peripheral keratitis.[1] In our instance, the patient had asymptomatic, plum-colored to hyperpigmented macules to papules on the dorsum of both hands and feet, along with a few nodules.

Acute febrile neutrophilic dermatosis, Dermatofibroma, erythema multiforme, granuloma annulare, granuloma faciale, multicentric reticulohistiocytosis, pyoderma gangrenosum, and xanthomas must be distinguished from the illness. The most helpful investigation for the identification of EED is a skin biopsy. Patients with EED frequently have increased erythrocyte sedimentation rates. Immunoelectrophoresis and antineutrophilic cytoplasmic Ig A antibodies may be beneficial.[1] EED is linked to a number of systemic diseases, including paraproteinemia, IgA monoclonal gammopathy, hairy cell leukemia, myeloproliferative disorders, and myelodysplasia. Aside from autoimmune diseases like systemic lupus erythematosus, celiac disease, rheumatoid arthritis, and Crohn's disease, other conditions linked to it include infections caused by HIV, group B streptococci, viral hepatitis, and syphilis, as well as cancers like B-cell lymphoma, multiple myeloma, and breast cancer.[1,7] Therefore, early diagnosis of EED is essential owing to the various associated conditions.

Because EED has a chronic and recurring course, treatment can be challenging. The most successful medication is dapsone, a sulfonamide antibiotic that inhibits neutrophil chemotaxis and function, but stopping therapy frequently results in relapse. NSAIDS, niacinamide, tetracyclines, chloroquine, colchicine, and plasmapheresis are some additional treatments. In mild instances, topical and intralesional corticosteroids might be helpful. In localized fibrotic lesions of EED, limited surgical excision can be helpful. If the underlying triggering factors are not controlled, the recurrence incidence of EED is significant, regardless of treatment.[1]

CONCLUSION

We have presented this case study for its rarity. EED is a rare chronic progressive dermatosis which can present with involvement of atypical sites such as palms which can mimic a number of other conditions. A high index of suspicion in such atypical presentations may facilitate early detection and treatment along with detection of other conditions associated with it.

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