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Case Report

Dermatology Section

Erythema Elevatum Diutinum: An Underdiagnosed Entity- A Case Report

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ABSTRACT

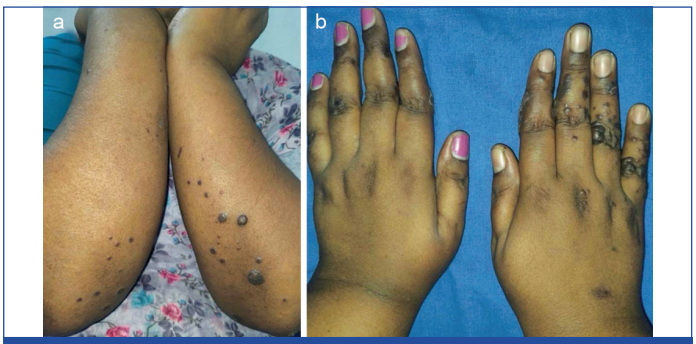
Erythema Elevatum Diutinum (EED), an uncommon form of chronic leukocytoclastic vasculitis presents as reddish-brown plaques and papulonodules primarily affecting the extensor surface. It mimics various cutaneous disorders and hence poses diagnostic difficulties. EED is predominantly seen in middle-aged individuals between fourth and sixth decade. EED can also have various extracutaneous manifestations such as arthralgia, scleritis, panuveitis, ulcerative keratitis and neuropathy indicating circulating immune complexes deposition in several organs. Thereby thorough clinical and systemic examination for early diagnosis and comprehensive management of both skin and extracutaneous findings are essential to prevent complications and further progression. The authors hereby report a case of 47-year-old female with long standing diabetes mellitus presenting with asymptomatic papulonodular lesions over the extensor surface with sensory abnormalities, highlighting the diagnostic dilemma and emphasising the importance of histopathological correlation in arriving at appropriate diagnosis.

Keywords: Leukocytoclasia, Neutrophils, Vasculitis

CASE REPORT

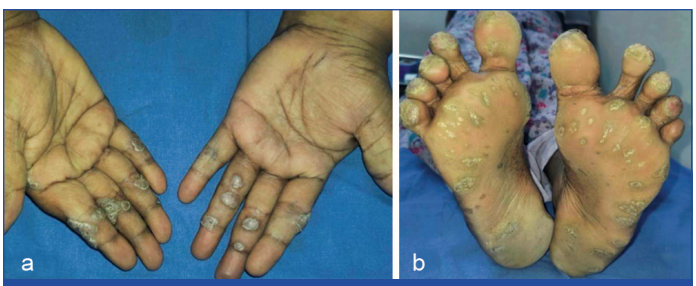
A 47-year-old female reported to the Dermatology Outpatient Department with the complaints of dark raised lesions over bilateral elbows as well as hands for the past two years associated with itching and burning sensation for the past one month. There was no history of sensory loss, atopy or drug intake before the occurrence of lesions. The patient was a known case of diabetes mellitus for the past 12 years and was on treatment.

Cutaneous examination revealed hyperpigmented nodules ranging from few millimetres to one centimetre over the extensor surface of forearm, elbows and dorsal aspect of fingers [Table/Fig-1a,b].



[Table/Fig-1]: a) Clinical image showing multiple hyperpigmented nodules over the extensor aspect of bilateral forearms and elbows; b) Clinical image showing multiple hyperpigmented nodules over dorsum of the bilateral hands.

Few nodules with scaling were present over the palms and soles [Table/Fig-2a,b].



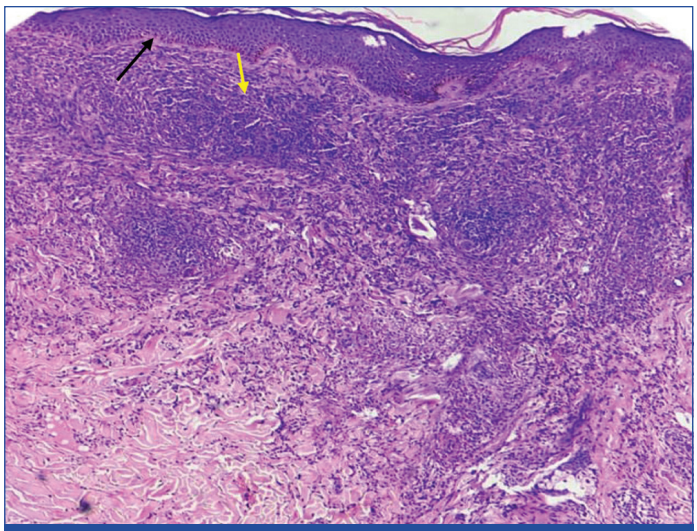
[Table/Fig-2]: a) Clinical image showing nodules with scaling over the palms; b) Clinical image showing nodules with scaling over the soles.

Nodules were soft in consistency and tender on palpation. On sensory examination, decreased sensation over bilateral feet was present. Peripheral Nervous System (PNS) examination was normal. Patient was diagnosed with axonal neuropathy of bilateral lower limbs through a nerve conduction study before the occurrence of skin lesions.

Clinically, diagnosis of EED, Hansen's disease and multicentric reticulohistiocytosis were considered.

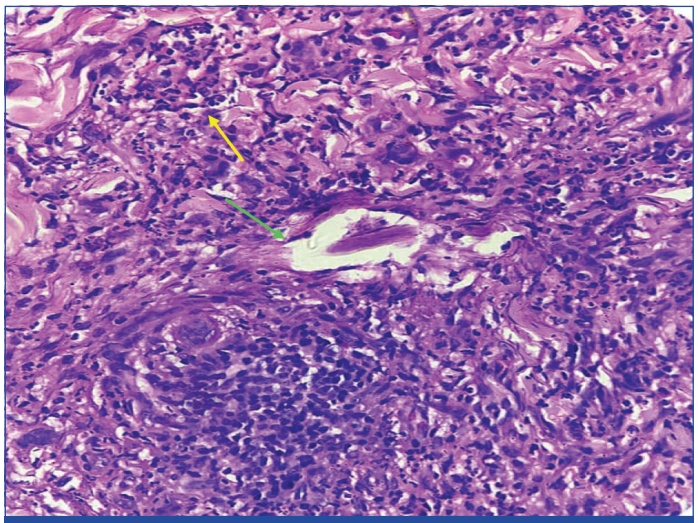
Standard investigations like complete blood count, liver function test, renal function test were within normal limits and viral markers like Hepatitis B surface Antigen (HBsAg), Integrated Counselling and Testing Centre (ICTC), Antibody to Hepatitis C Virus (HCV) were negative. Glycosylated haemoglobin (HbA1c) was 6.5% and post-prandial blood sugar levels were 277 mg/dL. Slit skin smear was negative for acid fast bacilli. Interferon Gamma Release Assay (IGRA) was negative and Ultrasonography (USG) abdomen was normal.

Histopathology showed epidermis with flattened rete ridges and superficial dermal infiltrate on low power view [Table/Fig-3].



[Table/Fig-3]: Histopathology showing epidermis with flattened rete ridges (black arrow) and superficial dermal infiltrate (yellow arrow) in low power view H&E, 10x.

In high power view, there was abundant neutrophilic infiltrate with a few eosinophils and minimal histiocytes around blood vessels with areas of leukocytoclasia and endothelial damage [Table/Fig-4].



[Table/Fig-4]: High power view showing abundant neutrophilic infiltrate (yellow arrow) in dermis with a few eosinophils and histiocytes along with leukocytoclasia and endothelial vessel wall damage (green arrow) (H&E, 40x).

A final diagnosis of EED was made based on clinicopathological findings, following which the patient was prescribed tablet Dapsone 100 mg once a day and intralesional steroid (Triamcinolone acetonide 40 mg/mL). The patient was lost to follow-up.

DISCUSSION

The EED, an uncommon vasculitis falls under neutrophilic dermatoses. EED was originally described in elderly men by Hutchinson in 1888 and in 1899 by Bury. The term "EED" was first coined by Henry Radcliffe-Crocker [1]. EED lesions typically present as firm, violaceous, red-brown plaques or papulo-nodules that are symmetrically distributed, particularly over the extensor surface of the elbow, knees, ankles, hands and fingers [2]. However, atypical sites such as truncal, retro-auricular, palmar and plantar areas may also be involved [3]. The present case highlights the classical symmetrical distribution of papulonodules over the extensors unlike the other atypical sites. EED is typically asymptomatic but may present with pruritus or burning as seen in the present case.

While EED primarily affects the skin, extracutaneous sites such as eyes and joints can also be affected. Conditions like peripheral keratitis, nodular scleritis, panuveitis, diffuse neuropathy were found in patients with EED [4]. Axonal neuropathy found in the present case was associated with uncontrolled diabetes mellitus as it was present before the onset of cutaneous lesions.

Various diseases that are found to be associated with EED include celiac disease, Human Immunodeficiency Virus (HIV), Hepatitis-B, tuberculosis, Paraproteinaemia, malignancies like non-Hodgkin's lymphoma, and connective tissue disorders [5]. Hence, serological workup like HBsAg, anti HCV, ICTC was done in the present case to rule out HIV, Hepatitis B,C and IGRA was done to rule out Tuberculosis. USG abdomen was also done to rule out internal malignancies.

On histopathological examination, predominant neutrophilic infiltrate with leukocytoclastic vasculitis can be appreciated in the early stages. In late stage of the disease extracellular cholesterolosis (cholesterol deposition in extracellular tissues) and fibrosis of the vessels with endothelial damage can be appreciated [6].

Multicentric reticulohistiocytosis, Hansen's disease, tuberous xanthoma, rheumatoid nodule, Sweet's syndrome can be considered as probable differentials for EED. The differentials which were considered in the current case were multicentric reticulohistiocytosis and Hansen's disease. In multicentric reticulohistiocytosis, the nodules will be associated with extracutaneous manifestations like anaemia, elevated Erythrocyte Sedimentation Rate (ESR), severe arthropathy and histopathology will show characteristic dermal infiltration of histiocytes, lymphocytes and a few multinucleated giant cells with eosinophilic ground glass cytoplasm, thus ruling out this condition in the current case [7]. In Hansen's disease, though the current case had nodules pointing towards Histoid Hansen's or lepromatous spectrum, staining for acid fast bacilli was negative and histological findings such as sheets of foamy histiocytes (Virchow cells), Grenz zone and the characteristic spindle shaped histiocytes of Histoid Hansen's were absent, ruling out these conditions. In case of rheumatoid nodule, rheumatoid factor will be positive and symptoms like arthritis will be present. In case of tuberous xanthoma, the characteristic nodules will be firm and yellowish occurring over the pressure sites and Fasting Lipid Profile (FLP) will be elevated. In Sweet's syndrome, there will be associated constitutional symptoms like fever and joint pain with plump nodules, plaques and pseudo-vesicles, and histology will show diffuse nodular and perivascular neutrophilic infiltrate without evidence of vasculitis [6,7].

The comparison of the findings of the present case with previous similar case reports is shown in [Table/Fig-5] [1,5,8,9].

Oral Dapsone is considered as the mainstay treatment. Topical or intralesional corticosteroids can be used to minimise the size of the lesions. In cases resistant to dapsone, niacinamide in combination

Variables/Authors	Sabooniha F [8] (2022)	Kumar NA et al., [1] (2023)	Qiao J et al., [5] (2024)	Ps K et al., [9] (2025)	Present case (2025)
Age and gender	32-year-old male	54-year-old female	61-year-old male	56-year-old female	47-year-old female
History	Tender raised skin lesions over the right hand for the past 8 years.	Multiple painful, dark coloured raised lesions over bilateral feet, arms and dorsum of fingers for the past 18 months	Painful skin lesions over the face, back, hands, elbows, buttocks for the past 15 years	Multiple painful swellings over the back and extremities for the past 2 years	Asymptomatic raised lesions over extensors for the past 2 years
Clinical features	Well defined firm red-brown papulonodule over the proximal interphalangeal joint of the third finger of the right hand	Multiple red brown coloured nodules were seen over the extensor aspect of the arms, forearms, dorsal aspect of hands, bilateral feet, over the elbow and knees.	Purple-red firm papules and plaques over the dorsal aspect of hands, elbows, thighs, gluteal region	Multiple tender, soft nodules over the posterior and medial aspect of the right elbow, dorsal and plantar aspect of the left foot	Hyperpigmented nodules over extensors of B/L elbows and knees
Diagnostic work-up	CRP, ESR, plasma electrophoresis, HBsAg, Anti HCV, HIV, Rheumatologic profile, biopsy	CBC, peripheral smear, ESR, CRP, Fasting lipid profile, HIV, X ray of the hands and feet, USG abdomen, skin biopsy, nerve conduction study	CBC, LFT, RFT, slit lamp examination, serum IgA levels, biopsy	CBC, LFT, FLP, RFT, ESR, CRP, biopsy, autoimmune workup	CBC, LFT, RFT, HBsAg, Anti HCV, ICTC, IGRA, USG abdomen, skin biopsy
Treatment	Oral dapsone 100 mg BD	Oral dapsone 100 mg OD	Triamcinolone 12 mg	Excision with split thickness skin grafting.	Oral dapsone 100 mg OD Intralesional Triamcinolone acetonide

[Table/Fig-5]: The comparison of the present case with other similar case reports from literature [1,5,8,9].
CBC: Complete blood count; LFT: Liver function test; RFT: Renal function test; IgA: Immunoglobulin A

with tetracycline, colchicine and even intermittent plasmapheresis can be considered. Newer treatment options include 5% topical dapsone preparation [10].

CONCLUSION(S)

The present case report highlights the typical clinical features, diagnostic workup and therapeutic options of EED in detail. EED is an under-diagnosed condition which can mimic other common dermatological conditions. Hence, detailed history, thorough clinical examination and diagnostic workup are required to arrive at accurate diagnosis and effective treatment, thereby preventing disease progression.

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