

# Erythema Elevatum Diutinum: A Rare Dermatosi s with an atypical Presentation

Mahesh Mathur<sup>1</sup>, Neha Thakur<sup>2</sup>, Supriya Paudel<sup>3\*</sup>, Sunil Jaiswal<sup>3</sup>, Srijana Maharjan<sup>3</sup> and Anjali Shrestha<sup>3</sup>.

<sup>1</sup>Head of the department, Department of Dermatology, College of Medical Sciences, Bharatpur, Nepal

<sup>2</sup>Lecturer, Department of Dermatology, College of Medical Sciences, Bharatpur, Nepal.

<sup>3</sup>MD resident, Department of Dermatology, College of Medical Sciences, Bharatpur, Nepal.

## \*Corresponding Author

Supriya Paudel, MD Resident in Department of dermatology College of Medical Sciences, Bharatpur, Nepal.

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## 1. Introduction

EED is a form of chronic cutaneous vasculitis of unknown underlying etiology [1]. EED was named by Henry Radcliff-Crocker and Williams in 1894 but discovered earlier in 1878 by Hutchinson. It presents clinically as asymptomatic, firm, erythematous to yellowish papules, nodules, or plaques involving the extensor surface of extremities often over joints [1]. Antigen-induced small vessel vasculitis is believed to be the cause. Various infections and systemic disorders like streptococcal throat infection, tuberculosis, HIV 1, HIV 2, Hepatitis B, Rheumatoid arthritis, ankylosing spondylitis, Sarcoidosis, Crohn's disease, Ulcerative colitis, Pyoderma gangrenosum, Paraproteinemia, Myelodysplasia, Kaposi's sarcoma have been reported to be

associated [1]. The clinical presentation of EED varies as the lesions progress leading to diagnostic difficulties [1].

## 2. Case Report

A 45-year-old male presented with asymptomatic, well-defined, periarticular papules gradually evolving into nodules predominantly on palms, dorsum of hands, and extensor surface of bilateral elbows for the last five years (Figures 1,2, and 3). Examination revealed well-defined, firm, yellowish to erythematous nodules measuring 0.5 to 5 cm in diameter. On palms, lesions were crusted and ulcerated with surrounding erythema (Figure 3). He visited multiple medical centers for his complaint without improvement.



**Figure 1:** Skin coloured nodules over dorsum of hands



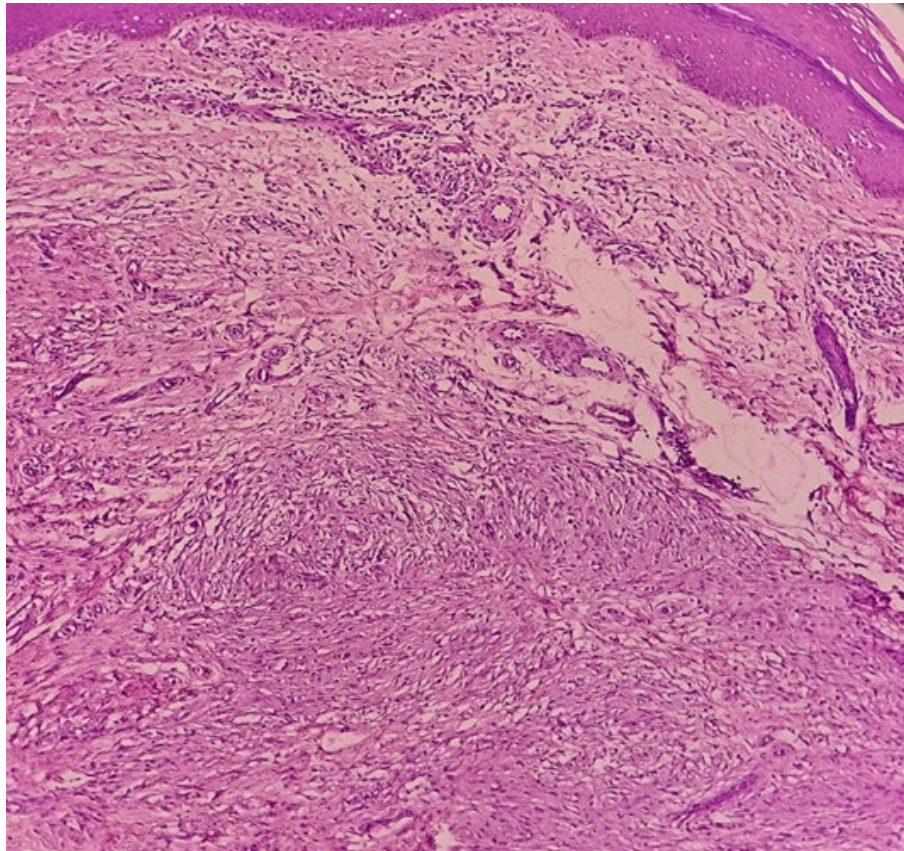
**Figure 2:** Erythematous to yellowish nodules over extensor aspect of elbows

Complete blood count, liver function test, renal function test, thyroid function test, serum electrolytes, anti-nuclear antibody (ANA), Rheumatoid factor, Anti-cyclic citrullinated peptide (anti-CCP), uric acid, and serological tests including HIV were within normal limit. An X-ray of his hands revealed osteophytic changes without joint involvement. Excisional biopsy of the nodule at the elbow revealed an epidermis with compact orthokeratosis, and a dermis showing leukocytoclastic vasculitis of small blood vessels along with well-circumscribed dermal nodule composed of dense cellular infiltrates. (Figure 4). Reticular dermis shows storiform fibrosis (Figure 4 and 5). Diagnosis of EED was made and he was started on dapsone. A good response was seen in the first month but he lost to follow-up.

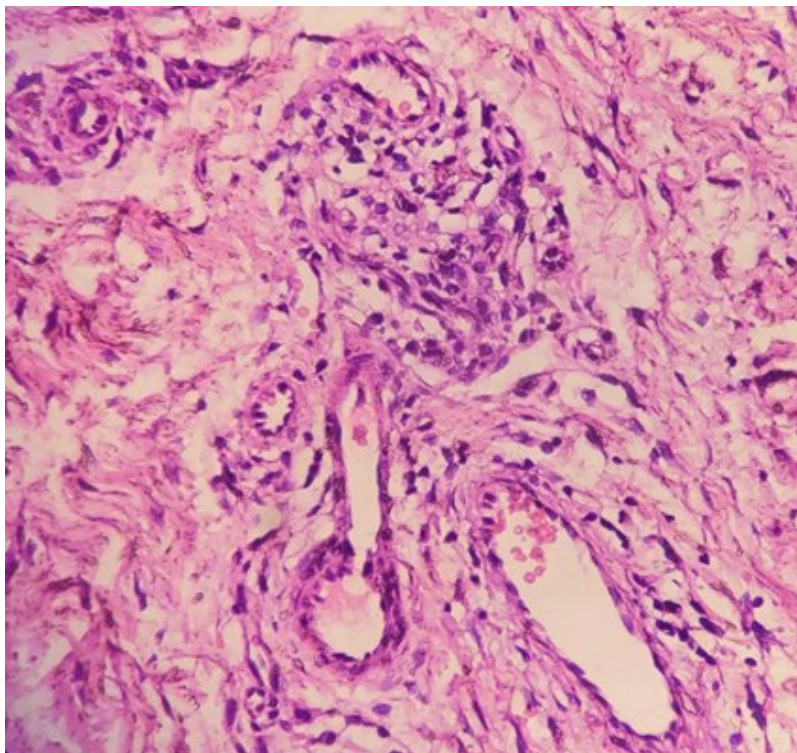


**Figure 3:** Ulcerated and crusted papules, nodules and plaques over palms





**Figure 4:** H and E (10x) shows storiform fibrosis in deep dermis with inflammatory infiltrates



**Figure 5:** Histopathology (H and E, 40x) showing dermal fibrosis and features of leukocytoclastic vasculitis

### 3. Discussion

EED is a rare dermatosis commonly affecting adults between 30 and 60 years of age with no gender or racial predilection. Bullous lesions may occur occasionally [2]. Palmar involvement, as seen in our case is rather an unusual manifestation of this rare disease. Ulcerated nodules on the palm are typical for HIV which is ruled out. The pathophysiology of EED is unknown. However, antigen-antibody immune complex deposition in cutaneous microvasculature is hypothesized as an inciting mechanism leading to chronic inflammation, fibrosis, and other sequelae of the disease [3]. EED usually follows a chronic course. Initial diagnosis is difficult due to the evolving nature of clinical and histologic findings. The histopathological examination at various stages shows nodular neutrophilic infiltrate, with features of leukocytoclastic vasculitis and subsequently, dermal fibrosis [2].

EED can be the first indicator of underlying comorbidity necessitating the correct diagnosis, screening, and management of associated conditions. None of the above conditions was present in our patient. The medical treatment of choice is dapsone in the early

stage [3]. In addition, anti-inflammatory agents like tetracyclines, niacinamide, non-steroidal anti-inflammatory drugs, methotrexate, chloroquine, colchicine, topical corticosteroids, plasmapheresis, and surgical intervention are other successful modalities of treatment reported [1]. Nevertheless, resolution of skin lesions may occur spontaneously after years.

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