

BRIEF ARTICLE

Erythema Elevatum Diutinum in Association with IgA Monoclonal Gammopathy of Undetermined Significance: A Case ReportKatherine De Jong, BSA¹, Mojahed Mohammad K Shalabi, MD², Dale Schaefer, MD, FAAD³¹ Texas A&M School of Medicine, Round Rock, TX² Department of Dermatology, Baylor Scott and White Medical Center, Temple, TX³ US Dermatology Partners Spicewood Springs, Austin, TX

ABSTRACT

Erythema Elevatum Diutinum (EED) is a distinctive form of chronic leukocytoclastic vasculitis characterized by red to brown papules, plaques, and nodules that favor the extensor aspect of the extremities. EED is a benign condition but may be associated with several systemic diseases, including hematologic disorders, infections, and autoimmune conditions. We present a rare case of a 61-year-old male with IgA monoclonal gammopathy of undetermined significance (MGUS) and a 17-year history of EED that improved significantly with dapsone treatment.

INTRODUCTION

Erythema Elevatum Diutinum (EED) is a rare form of chronic leukocytoclastic vasculitis, which presents as firm, symmetrical red to brown papules or nodules on the skin.¹ Leukocytoclastic vasculitis is characterized by small vessel inflammation of the dermal capillaries and venules.² The exact mechanism of EED is unknown, but it is thought to be due to immune-complex deposition with resultant complement fixation and inflammation of dermal blood vessels. EED has been associated with many systemic diseases, including hematologic disorders, infections, and autoimmune conditions.³ This case report includes a 61-year-old male with MGUS and a 17-year history of undiagnosed EED that significantly improved with dapsone treatment.

CASE REPORT

A 61-year-old man presented with a 17-year history of multiple, itchy, painful, reddish to violaceous lesions present on the face, trunk, upper and lower extremities and a 3-year history of similar lesions on the bilateral palms, soles, dorsal hands, and external ears. The lesions initially appeared as two mildly pruritic, painless, erythematous plaques over the bilateral buttocks. Mycosis fungoides was suspected and the patient was treated in India with oral prednisolone for a week which provided temporary relief. Nevertheless, the lesions recurred with increased size and associated symptoms such as itching, pain, and burning sensation. The lesions progressed to involve the extensor surface of the bilateral upper arm, bilateral knees and elbows, palms and soles, external ears, trunk, and face. Over a time span of 15 years, he had undergone multiple

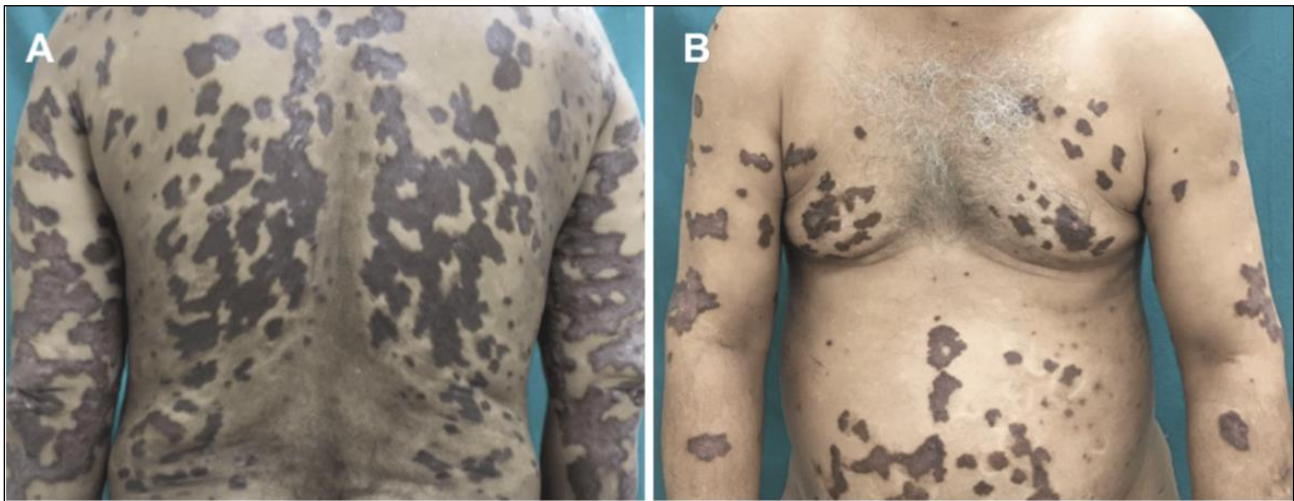


Figure 1. EED prior to Dapsone treatment. (A) Multiple erythematous to violaceous plaques over posterior trunk and upper extremities. (B) Multiple erythematous to violaceous plaques over anterior trunk and upper extremities.

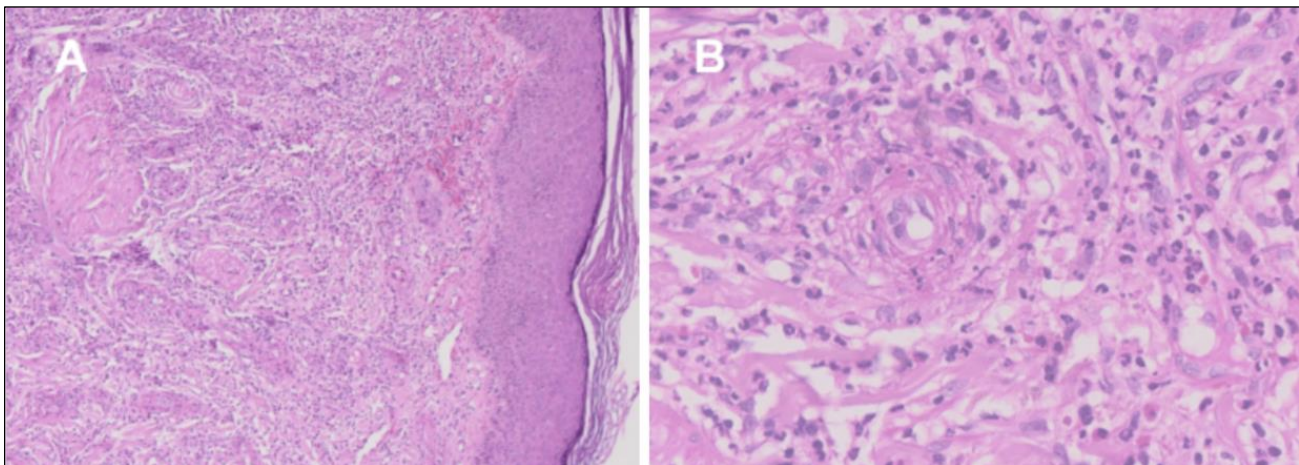


Figure 2. Leukocytoclastic vasculitis. (A) Polymorphous infiltrate includes neutrophils, lymphocytes, histiocytes, plasma cells and eosinophils. (B) Perivascular fibrosis in onion skinning pattern.

dermatological and alternative medicine evaluations and was misdiagnosed with psoriasis vulgaris. He was given oral methotrexate (15mg/week) which he had been inconsistently taking for the past 15 years with little to no relief. Some lesions regressed but left a trace of post-inflammatory hyperpigmentation and atrophy. He had multiple episodes of recurrence and development of new lesions while on the medication which prompted him

to seek different medical care. Upon re-examination a multitude of infectious diseases, inflammatory diseases, autoimmune diseases, vascular diseases, and malignancy were considered in the work-up.

Clinical Findings

On skin examination, there were multiple, well-defined, discrete to confluent,

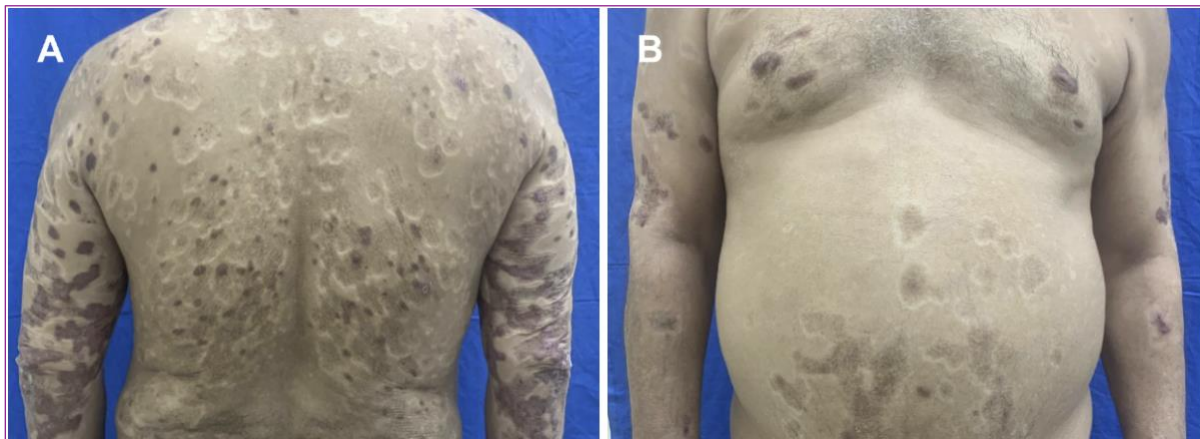


Figure 3. EED 24 months following Dapsone treatment. (A) Resolution of lesions on posterior trunk with scarring. (B) Resolution of lesions on anterior trunk with scarring.

erythematous to violaceous plaques with irregular margins on the upper and lower extremities, neck, trunk, and bilateral ears (**Figure 1**). In addition, there were multiple hyperkeratotic, scaly plaques studded with firm papulonodules present over the bilateral dorsal of the hands, palms, and soles. Laboratory investigations showed CBC, PBF, ESR, LFT, RST, RFT, serum electrolytes, Antistreptolysin O titer, Serum ACE levels, and urine microscopy were in normal ranges. Mantoux test, Viral serology (HBsAg, HCV, HIV), VDRL, ANA /ANCA / RA factor, slit skin smear, Covid 19 RT PCR were negative as well. Chest radiograph showed no abnormalities. His HbA1c level was 6.5%.

Diagnostic Assessment

The differential diagnosis for the clinical presentation included disseminated discoid lupus erythematosus, lupus erythematosus and lichen planus overlap syndrome, mycosis fungoides, and erythema elevatum diutinum. Skin biopsy was taken from a violaceous plaque and histopathology revealed leukocytoclastic vasculitis with polymorphous infiltrate, including neutrophils, lymphocytes, histiocytes, plasma cells, and eosinophils (**Figure 2A**).

Perivascular fibrosis in an “onion skinning” pattern was also present (**Figure 2B**). The biopsy confirmed the diagnosis of EED, and the patient was referred to clinical hematology for further workup due to suspicion of monoclonal gammopathy.

Upon further hematologic investigation, a serum protein electrophoresis revealed an M band present (0.74 g/dL), and immunofixation revealed IgA lambda. These results confirmed the diagnosis of monoclonal gammopathy of undetermined significance (MGUS).

Therapeutic Interventions

The patient was started on dapsone (100 mg once daily) treatment. Individual, highly symptomatic lesions also responded very well to intralesional Kenalog. After 6 months of dapsone treatment, resolution of the lesions was noted, as well as no new lesions forming. After 24 months of regular treatment, complete resolution of lesions was noted. The lesions healed with hypopigmented, slightly depressed scars (**Figure 3**).

DISCUSSION

Erythema elevatum diutinum is a rare, distinctive form of vasculitis that often peaks in the sixth decade of life. Lesions preferentially appear in the extensor regions and present as firm, brownish to red-colored papules, plaques, or nodules. Atypical sites, such as truncal, retroauricular, plantar, and palmar regions, have also been shown to occur. The lesions, while mostly asymptomatic, can present with pain and pruritus.¹

While the pathogenesis is not fully understood, EED is thought to be due to immune-complex deposition in dermal vasculature that activates the complement cascade.² Histologically, EED is characterized as leukocytoclastic vasculitis with polymorphous infiltrate, including neutrophils, lymphocytes, histiocytes, plasma cells, and eosinophils. In the later stages of the disease, perivascular fibrosis can be seen.³ EED is a benign condition but is known to be associated with several systemic diseases, including hematologic, rheumatologic, and infectious causes.

This present case was associated with IgA monoclonal gammopathy of undetermined significance (MGUS). There have been reports of monoclonal IgA gammopathy and IgA myeloma in association with EED since 1977.⁴ A study by Yiannias et al. found that 6 out of 13 patients with EED had hematologic abnormalities. Of the six patients with hematologic abnormalities, IgA clonal gammopathies were found to be the most frequently associated hematologic condition (4 patients).

Following the diagnosis of EED, the patient was started on Dapsone 100mg/day. After six months of continuous dapsone treatment,

resolution of the lesions was noted, and no new lesions were forming. After 24 months of regular treatment, complete resolution of lesions was noted. The lesions healed with hypopigmented, slightly depressed scars.

CONCLUSION

We report this rare case of chronic dermatitis presenting with well-defined, violaceous infiltrated plaques with hyperpigmented borders distributed on the upper extremities, lower extremities, neck, trunk, bilateral hands, and bilateral ears. Histology showed leukocytoclastic vasculitis with classic perivascular “onion skinning” fibrosis, confirming the diagnosis of EED. The patient was subsequently treated with dapsone 100 mg once daily and complete resolution of lesions was noted. Dapsone is the primary treatment for EED.

Conflict of Interest Disclosures: None

Funding: None

Corresponding Author:

Katherine De Jong
803 West 30th Street, Austin, TX, 78705
Email: katiedejong@tamu.edu

References:

1. Lawrence E. Gibson, Rokea A: el-Azhary, Erythema elevatum diutinum, Clinics in Dermatology, Volume . 18:295-299. 10.1016/s0738-081x(99)00120-0
2. Rashdan H, Schafer H, Lundgren AD, O'Connor K, Keeling B: Transient Erythema Elevatum Diutinum Associated With HIV Viremia. Cureus. 2023;15(6):e40858. 10.7759/cureus.40858
3. Patnala GP, Sunandini AP, Rayavarapu R, Yandapalli PS: Erythema elevatum diutinum in association with IgA monoclonal gammopathy: A rare case report. Indian Dermatol Online J. 2016;7(3):300-303. 10.4103/2229-5178.185472
4. James A. Yiannias, MD, Rokea A. El-Azhary, MD, Lawrence E. Gibson, MD : Erythema

July 2024 Volume 8 Issue 4

elevatum diutinum: A clinical and histopathologic study of 13 patients. Journal of the American Academy of Dermatology. 1992, 26:38-44. 10.1016/0190-9622(92)70003-X