A Case of Erythema Elevatum DiutinumAssociated with IgA Monoclonal Gammopathy and Multiple Myeloma

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Introduction

- Erythema Elevatum Diutinum (EED) is a rare type of chronic leukocytoclastic vasculitis characterized by symmetric violaceous, red, or yellowish papules, plaques, or nodules.
- These are typically seen on the extensor surfaces of the body, mostly involving the extremities, without any systemic vasculopathy.
- Usually, it presents at middle age and is associated with arthralgia, peripheral ulcerative keratitis, and pulmonary infiltrates.
- EED has been reportedly associated with multiple systemic diseases:
  - Recurrent bacterial infections (streptococcal) and viral infections (HBV, HHV-6, and HIV).
  - Myelodysplastic syndrome, lymphoma, IgA monoclonal gammopathy, multiple myeloma.
  - Rheumatoid arthritis, systemic lupus erythematosus, Wegener's granulomatosis, and cryoglobulinemia.
  - Celiac disease, ulcerative colitis, Crohn's disease.
  - Pyoderma gangrenosum.
  - Hyperimmunoglobulinema D syndrome.
  - Relapsing polychondritis.
  - POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin lesions).
- EED is usually treated with dapsone and corticosteroids.

Case Description

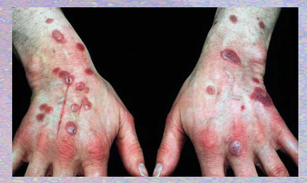
History

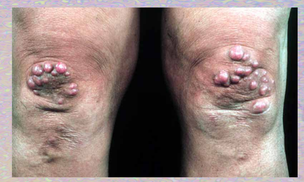
- C.C. is now a 41 years old Hispanic female who initially presented to the emergency department in 2006 after she developed an itchy, burning, purple rash (papules, plaques, and nodules) on her elbows, knees and posterior thighs.
- Appearance of these lesions was preceded by months of widespread joints pain.
- Patient was referred to a dermatologist who suspected EED and placed the patient on dapsone.
- The diagnosis was confirmed with a biopsy of the skin lesions.
- Patient responded dramatically to the dapsone with rapid resolution of existing rash. However, she had multiple relapses every time she stopped taking the medication.

Physical Exam

Investigational studies

- Given the potential association of EED with multiple systemic diseases, the patient had an extensive work up.
- All serologic testing were negative, including ANA, RF, CCP, tTG IgA Ab, C- and P-ANCA, and complement levels.
- Other negative work up includes TSH, G6PD, ASO, HIV, and hepatitis panel.
- Iron studies were consistent with iron deficiency anemia.
- Serum protein electrophoresis and immunofixation were initially negative. However, a repeated SPEP couple years later showed evidence of IgA kappa monoclonal protein (monoclonal gammopathy).
- Bone marrow biopsy showed normocellular marrow involved by a clonal plasma cell disorder.
- Skeletal survey showed no suspicious lesions.





Patient Progress

- The hematologist diagnosed the patient with asymptomatic smoldering myeloma and continues to follow her closely.
- No treatment at this time

Discussion

- We report the development of IgA monoclonal gammopathy and later multiple myeloma in a patient with chronic EED on dapsone therapy.
- EED is an uncommon disease and its association with systemic disorders, including neoplastic diseases, is exceedingly rare.
- This case emphasizes the need for extensive work up and close surveillance of patients with EED to diagnose life-threatening systemic diseases early enough to have a chance to successfully treat the associated morbidities.