

DERMATOPATHOLOGY UNIT

UPP | Department of Dermatology

UPMC Dermatopathology "Case of the Month" Presentations

UPP - Department of Dermatology, Dermatopathology Unit

5230 Centre Avenue (412) 623-2614

Pittsburgh, PA 15232 (412) 682-6450 FAX

Jason G. Whalen, MD; Matthew Zirwas, MD; Hina Sheikh, MD; Drazen M. Jukic, MD

JANUARY 2006 CASE OF THE MONTH

CLINICAL FINDINGS

Clinical History

A.D. was a 57 year old white male who presented to our clinic with a 6 month history of an asymptomatic diffuse rash that was primarily distributed on his chest, abdomen, buttocks, and lower extremities. There was no pain or pruritus experienced in these lesions, and he did not recall any bite or trauma to his skin. The lesions did not migrate, and he continued to get new ones as others faded. He had not tried any treatments as of the date we saw him. All of his age-appropriate malignancy screening was performed within the last year and they were all normal. A.D.'s past medical history included hypertension and hypercholesterolemia. His medications included Lipitor (statin) which he started about 8 months ago, and Micardis (angiotensin II receptor (type AT1) antagonist) for approximately 2 years. He had no known allergies. No other family members experienced similar rashes. He was a non-smoker and non-drinker. Review of systems was entirely negative. The following labs and results ordered by his primary care physician are as follows: ANA- Negative, WBC- 5.9(Normal Diff), Hgb-15.1, Hct-43.8, Plts-224, ESR-5, Lyme titers- Negative.

Review of systems was unremarkable. She had no significant past medical history, no family history of similar lesions or autoimmune disease, and was not taking any medications.

Physical Exam

On exam, he had several large (5-20 cm), annular, non-scaly, pink plaques with central clearing scattered on his chest, abdomen, buttocks and lower extremities (Figures 1 & 2).

Histopathology

Two 4 mm punch biopsies were obtained from the edges of 2 different lesions- 1)right abdomen and 2)right thigh (Figures 3 & 4). In both specimens, superficial and deep perivascular neutrophilic infiltrates are present that also distribute around periadnexal structures. Eosinophils and flame figures are numerous and distributed interstitially as well as perivascularly. Some increase in CD 68 positive macrophages is also seen, and multiple mast cells are outlined the tryptase.

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Figures & Images

- Figures 1- Clinical Photo
- Figures 2- Clinical Photo
- Figures 3- Histopathology Photo- Low Power
- Figures 4- Histopathology Photo- High Power

1. Click on the Figure number you wish to review.
2. Click on the image to enlarge



Fig1



Fig 2

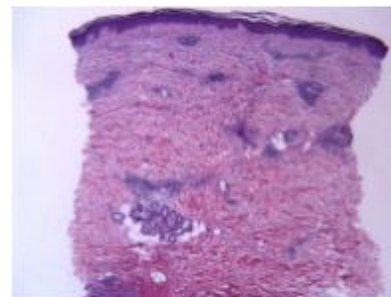


Fig 3

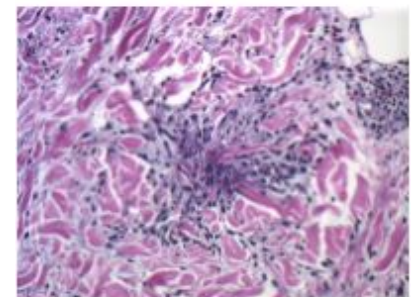


Fig 4

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DISCUSSION & DIAGNOSIS

Diagnosis

Acrokeratoelastoidosis of Costa

Discussion

The interstitial granulomatous drug reaction (IGDR) is a relatively new, rare and evolving topic in dermatology. The first papers on this condition were published in 1998 and only a few have been published since.

IGDR is thought to resemble the condition "interstitial granulomatous dermatitis with plaques" or "interstitial granulomatous dermatitis with arthritis" described by Ackerman et al. both clinically and histopathologically. Most patients have reproducible clinical findings including erythematous-to-violaceous, nonpruritic plaques, often with an annular pattern, predominantly involving inner aspects of the arms, medial thighs and intertriginous areas. The most common implicated drug classes include calcium channel blockers, angiotensin converting enzyme inhibitors, beta-blockers, and lipid-lowering agents, although other drugs have been reported.

The pathogenesis for most medications remains unknown, however, all of the implicated agents are known to have immune-dysregulating properties. In regards to the ACE inhibitor class, they down-regulate and alter type-specific collagen production by decreasing angiotensin II. The most frequent clinical differential diagnoses included on the pathology report by the clinician are cutaneous T-cell lymphoma, erythema annulare centrifigum (EAC), erythema gyratum repens, erythema migrans, granuloma annulare, erythema nodosum, and lupus erythematosus. The pathology of IGDR most commonly shows a dense, diffuse histiocytic infiltrate distributed interstitially and in palisaded array within the reticular dermis. Eosinophils and some neutrophils are usually scattered throughout the infiltrate. In some areas, surrounded by histiocytes, are thick collagen bundles associated with basophilic nuclear debris or "flame figures".

The only known treatment is to stop the offending drug. The time course to resolution of previous published cases has been anywhere from one week to one year. The patient's primary care physician stopped his Lipitor and changed to his Micardis to hydrochlorothiazide. There was slight lightening of his lesions at a follow-up 1 week later for suture removal.

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