Recurrent erythematous patch on the upper arm

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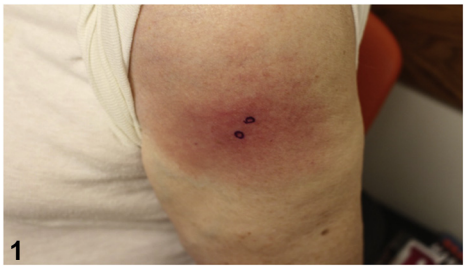

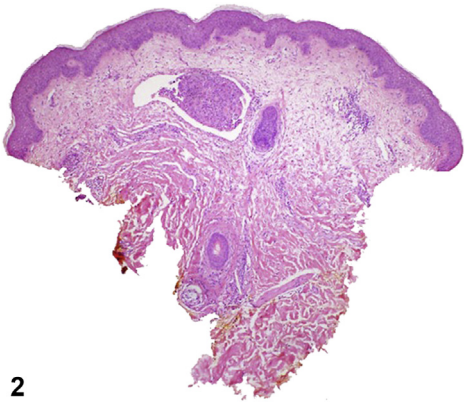

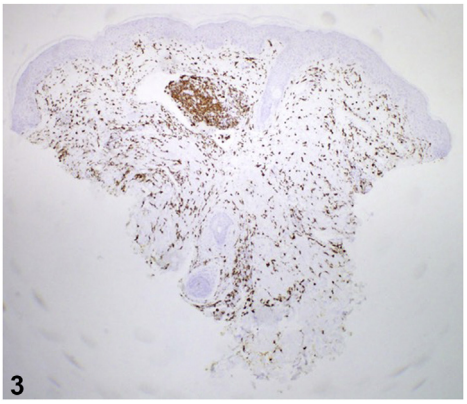

An 89-year-old man presented with a 1-year history of an intermittently warm, erythematous patch, associated low-grade fevers, and left upper arm tenderness and weakness. The patient was previously hospitalized and treated for cellulitis with intravenous vancomycin and piperacillin/tazobactam with no improvement. Physical examination found a temperature of 100.4°F and a large, ill-defined, nonindurated, warm, and erythematous patch on the left lateral upper arm (Fig 1). A punch biopsy found demonstrated mononuclear cells with pale cytoplasm without obvious atypia within dilated superficial vessels (Fig 2). These cells were positive for a CD163 immunostain (Fig 3).

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Question 1: What is the most likely diagnosis?

A. Angiosarcoma

B. Intralymphatic histiocytosis (IH)

C. Intravascular large B-cell lymphoma

D. Lupus erythematosus panniculitis

E. Cellulitis

Answers:

A. Angiosarcoma — Incorrect. Although common in elderly patients, angiosarcoma lesions are typically purpuric patches commonly on the central face, forehead, or scalp. More advanced lesions are violaceous nodules or indurated plaques that may ulcerate. Histopathology shows an anastomosing network of blood vessels that are lined with endothelial cells with variable degrees of nuclear atypia and infiltrate collagen bundles and subcutaneous fat.[1]

B. IH — Correct. The pathogenesis of IH remains unknown. Clinically, lesions vary but commonly present with ill-defined erythematous patches or plaques with livedo-reticularis—like lesions located on the upper or lower extremities. Histopathologically, IH is characterized by lymphangiectasia with intraluminal aggregates of histiocytes, which is confirmed by immunohistochemistry staining.[2] CD163, a histiocytic marker, was positive in our case.

C. Intravascular large B-cell lymphoma — Incorrect. Although intravascular large B-cell lymphoma can present as an erythematous or violaceous patch or plaque, it commonly affects the low extremities and trunk. Moreover, it often has systemic involvement including the central nervous system, and lesional cells demonstrate evidence of B-cell lineage.[1]

D. Lupus erythematosus panniculitis — Incorrect. Although Lupus erythematosus panniculitis commonly affects the arms, it is characterized by tender subcutaneous nodules and plaques with variable overlying epidermal changes such as scaling, follicular plugging, dyspigmentation, telangiectasias, atrophy, and scarring. Histopathology shows a lobular lymphoplasmacytic infiltrate in the subcutaneous fat.[1]

E. Cellulitis — Incorrect. Cellulitis often presents with fevers, ill-defined erythema, edema, and warmth; however, improvement should have resulted from antibiotic administration. Histopathology shows an inflammatory infiltrate of lymphocytes and neutrophils throughout the dermis and often extending into subcutaneous fat.[1]

Question 2: Which of the following has been associated with this diagnosis?

A. Rheumatoid arthritis (RA)

B. Intravenous drug abuse

C. Kaposi sarcoma

D. Systemic sclerosis

E. Sulfonamide antibiotics

Answers:

A. RA — Correct. There is a frequent association between IH and RA, although the clinical course of cutaneous lesions involved in IH does not parallel the course of RA.[2] There are also several cases of cutaneous IH found on mastectomy surgical scars of previous breast carcinoma,[2] near the joints of metal implants,[2,3] and in osteoarthritis.[3] It is hypothesized that these associated conditions result in chronic inflammation at the joint and subsequent development of lymph stasis and lymphangiectasia, which leads to poor antigen clearance, local inflammatory response, and subsequent aggregation of histiocytes.[2]

B. Intravenous drug abuse — Incorrect. To our knowledge, there has been no association between IV drug abuse and IH reported in the literature.

C. Kaposi sarcoma — Incorrect. To our knowledge, there has been no association between Kaposi sarcoma and IH reported in the literature.

D. Systemic sclerosis — Incorrect. To our knowledge, there has been no association between Systemic sclerosis and IH reported in the literature.

E. Sulfonamide antibiotics — Incorrect. To our knowledge, there has been no association between the administration of sulfonamide antibiotics and IH reported in the literature.

Question 3: Which of the following treatments is found to be therapeutically effective against IH in the setting of RA?

A. Local radiotherapy

B. Topical corticosteroids

C. Cyclophosphamide

D. Infliximab

E. Amoxicillin plus acetylsalicylic acid

Answers:

A. Local radiotherapy — Incorrect. Partial responses with relapses were seen with the use of local radiotherapy.[2]

B. Topical corticosteroids — Incorrect. Partial responses with relapses were seen with the use of topical steroids.[2] Intralesional triamcinolone and pressure bandage were found to be effective for IH associated with joint replacement but with no history of RA.[4]

C. Cyclophosphamide — Incorrect. Partial responses with relapses were seen with the use of cyclophosphamide.[2]

D. Infliximab — Correct. Cutaneous IH lesions and associated arthralgia secondary to RA significantly improved in a 66-year-old woman after administration of 3-mg/kg dose of intravenous infliximab. Given improvement with a tumor necrosis factor inhibitor, it is believed that tumor necrosis factor may play a crucial role in the pathogenesis of IH.[5]

E. Amoxicillin plus acetylsalicylic acid — Incorrect. Partial responses with relapses were found with the use of amoxicillin plus acetylsalicylic acid.[2]

Abbreviations used:

IH: intralymphatic histiocytosis
RA: rheumatoid arthritis

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