A 36-year-old African American woman with anemia, multiple joint arthralgias, synovitis of the hand, and chronic pain presented with a 6-month progressive nodular eruption without induration on the arms, legs, and trunk (Fig 1). Review of symptoms was negative without any exposure to radiotherapy or gadolinium contrast. Laboratory evaluation revealed positive antibody titers: antinuclear antibody, >1:2560; anti–double-stranded DNA, >300 IU/mL; Sjögren syndrome-A (RO) 1.2 AI; and anti-Smith; ribonucleoprotein IgG; and small nuclear ribonucleoprotein/ribonucleoprotein, >8.0 AI. Creatinine and blood urea nitrogen levels were 1.45 mg/dL and 24 mg/dL, respectively. Scleroderma (Scl-70) antibody and anti-JO 1 antibody IgG were negative, and thyroid function tests, serum protein electrophoresis, immunofixation, and serum-free light chain were within normal limits. A biopsy specimen was obtained (Fig 2).

Question 1: What is the most likely diagnosis?

A. Scleromyxedema (papular mucinosis)
B. Systemic sclerosis (scleroderma)
C. Nephrogenic systemic fibrosis (nephrogenic fibrosing dermopathy)
D. Cutaneous lupus mucinosis (papulonodular mucinosis)
E. Subacute cutaneous lupus erythematosus

Answers:

A. Scleromyxedema (papular mucinosis) — Incorrect. Papules in scleromyxedema tend to be symmetric, widespread, and waxy, commonly arranged in linear groups with progressive sclerodermoid induration. Pertinent negatives further decrease the likelihood of scleromyxedema, including normal thyroid function and normal protein immunoelectrophoresis, immunofixation, and serum-free light chains. Scleromyxedema and cutaneous lupus mucinosis may have similar clinical presentations; however, in the context of the serologic findings, this is not scleromyxedema.

B. Systemic sclerosis (scleroderma) — Incorrect. Papular and nodular mucinosis may also present as findings of early systemic sclerosis1; however, our patient had a normal scleroderma (Scl-70) antibody, no clinical evidence of sclerosis, and a negative review of symptoms for systemic sclerosis, including esophageal dysmotility, Raynaud phenomenon, and telangiectasias.

C. Nephrogenic systemic fibrosis (nephrogenic fibrosing dermopathy) — Incorrect. The patient had no history of radiologic imaging with gadolinium-based contrast or significant renal insufficiency. antinuclear, anticentromere, and anti-Scl-70 antibodies were negative, and serum protein electrophoresis, immunofixation, and light chain were insignificant.

D. Cutaneous lupus mucinosis (papulonodular mucinosis) — Correct. Nodular cutaneous lupus mucinosis is a rare manifestation of systemic lupus erythematosus that presents with nodules on the trunk and upper and lower extremities.2 Histology typically shows abundant mucin deposition dispersed among collagen bundles within the dermis. On the basis of the combination of clinical, pathologic, and serologic findings, she was diagnosed with systemic lupus erythematosus and associated nodular cutaneous lupus mucinosis.

E. Subacute cutaneous lupus erythematosus — Incorrect. Subacute cutaneous lupus erythematosus is a widespread nonscarring photosensitive rash presenting as annular plaques with raised borders and central clearing. Clinically, the quantity of mucin is not sufficient to produce detectable lesions. Histologically, there is epidermal involvement and vacuolar degeneration of the dermoeipidermal junction.3

Question 2: Which of the following histologic findings corresponds to nodular cutaneous lupus mucinosis?

A. Deposition of mucin that is surrounded by collagen bundles within the dermis

B. Deposition of mucin along with fibrosis and fibroblast proliferation

C. Thick bundles of collagen in the dermis and presence of inflammatory T cells

D. Thick bundles of collagen in the dermis along with mucin deposition and CD34+ fibrocytes. No significant presence of inflammatory cells

E. Scattered deposits of calcium in the dermis

Answers:

A. Deposition of mucin that is surrounded by collagen bundles within the dermis — Correct. Deposition of mucin that is surrounded by collagen bundles within the dermis is the classic histologic hallmark of nodular cutaneous lupus mucinosis.1,4

B. Deposition of mucin along with fibrosis and fibroblast proliferation — Incorrect. Deposition of mucin along with fibrosis and fibroblast proliferation represents scleromyxedema.

C. Thick bundles of collagen in the dermis and presence of inflammatory T cells — Incorrect. Thick bundles of collagen in the dermis and the presence of inflammatory T cells represent the early stages of systemic sclerosis.

D. Thick bundles of collagen in the dermis along with mucin deposition and CD34+ fibrocytes. No significant presence of inflammatory cells — Incorrect. Thick bundles of collagen in the dermis along with mucin deposition and CD34+ fibrocytes (and no significant inflammatory infiltrate) are classic hallmarks for nephrogenic systemic fibrosis.

E. Scattered deposits of calcium in the dermis — Incorrect. Scattered deposits of calcium in the dermis are observed in calcinosis cutis.

Question 3: What is the best first-line treatment for nodular cutaneous lupus mucinosis?

A. Sun avoidance and protection

B. Topical steroids

C. Antimalarials
D. Immunosuppressants

E. Isotretinoin

Answers:

A. Sun avoidance and protection — Incorrect. Sun avoidance and protection should be discussed with all patients with cutaneous lupus erythematosus; however, these are only preventive measures.

B. Topical steroids — Incorrect. It is unlikely that topical steroids alone will treat the nodules and papules associated with cutaneous lupus erythematosus; however, they can be combined with systemic therapies.

C. Antimalarials — Correct. Antimalarials are the first-line systemic therapy for the prevention and treatment of cutaneous and systemic lupus erythematosus. In this patient, the papulonodular lesions cleared after 2 months on hydroxychloroquine (200 mg twice a day).

D. Immunosuppressants — Incorrect. Immunosuppressants, including mycophenolate mofetil, azathioprine, methotrexate, and prednisone, are effective second-line systemic therapies selected in concordance with comorbidities.

E. Isotretinoin — Incorrect. Isotretinoin is used for the treatment of acne.

Conflicts of interest

None disclosed.

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