A hyperpigmented plaque in a female patient

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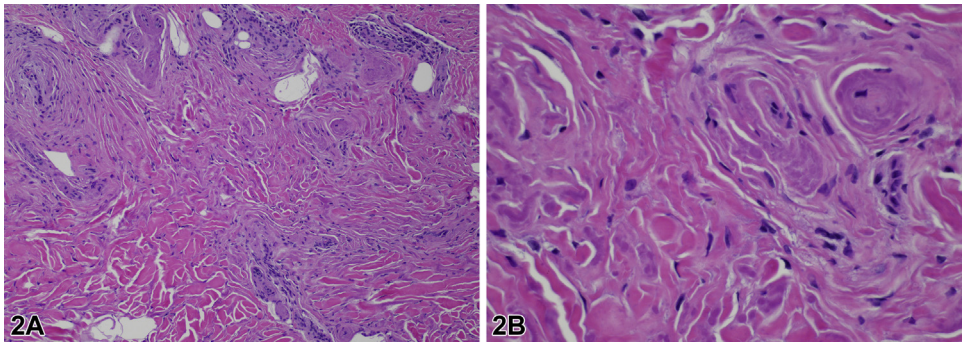

INTRODUCTION

A 59-year-old African American female with a past medical history of hypertension, depression, and arthritis presented for initial evaluation of a lesion on the upper portion of her back (Fig 1). The lesion had been slowly growing over the past few years and was occasionally pruritic. On examination, it appeared to be a waxy, firm, hyperpigmented, and painless plaque. She denied any history of keloids as well as prior trauma to the area, fever, chills, or unexplained weight loss. Upon biopsy of the lesion, histopathology revealed deposition of an extracellular pink hyaline material as well as an infiltrate of perivascular plasma cells (Fig 2).

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Question 1: What is the most likely diagnosis?

A. Seborrheic keratosis

B. Keloid

C. Dermatofibrosarcoma protuberans

D. Macular amyloidosis

E. Nodular cutaneous amyloidosis

Answers:

A. Incorrect - Seborrheic keratosis is a common skin finding and can present as a darkly pigmented plaque. The histopathology of seborrheic keratosis would show papillomatosis, acanthosis, and associated keratin-filled "horned pseudocysts."

B. Incorrect - Keloids are skin lesions that can present as nontender, slow-growing skin plaques. However, our patient's lesion was not associated with any previous trauma, which is typically seen in the case of keloids. Also, the histopathology of a keloid would exhibit proliferation of fibroblasts and collagen, which was not seen in the present case.

C. Incorrect - Dermatofibrosarcoma protuberans is a rare, locally aggressive soft-tissue sarcoma that can present as a growing hyperpigmented skin plaque. Its histopathology features monomorphic spindle cells arranged in an irregularly whorled or storiform pattern, often said to resemble a "straw mat," which was not observed in this case.[1]

D. Incorrect - Macular amyloidosis is a subtype of primary cutaneous amyloidosis that typically presents in the interscapular area. On histopathology, sparse pink deposits are limited to the papillary dermis and may mimic normal skin.[2]

E. Correct - Nodular cutaneous amyloidosis is the rarest subtype of primary cutaneous amyloidosis.[2] The histopathology of a specimen from this lesion featuring extracellular pink hyaline material is indicative of amyloid deposition. Plasma cell infiltrate and subcuticular involvement indicate nodular cutaneous amyloidosis rather than other subtypes of cutaneous amyloidosis.[2]

Question 2: What are the amyloid plaques seen in the histopathology most likely made of?

A. Immunoglobulin light chains

B. Serum amyloid protein A

C. Keratin derivatives

D. Amyloid precursor protein

E. Beta-2-microglobulin protein

Answers:

A. Correct - Nodular cutaneous amyloidosis is caused by the deposition of light-chain amyloid made up of immunoglobulin light chains, which are thought to be synthesized by local monoclonal plasma cells in the skin.[2,3]

B. Incorrect - Serum amyloid protein A makes up AA amyloid, which is characteristically seen in systemic amyloidosis resulting from chronic inflammatory conditions like rheumatoid arthritis.[2,3]

C. Incorrect - Keratin derivatives make up the amyloid deposits found in lichen amyloidosis and macular amyloidosis.[2,3]

D. Incorrect - Amyloid precursor protein is synthesized in the central nervous system and is responsible for dementia, which is associated with Alzheimer disease; it plays no role in nodular cutaneous amyloidosis.

E. Incorrect - Beta-2-microglobulin protein constitutes the amyloid deposits that are classically seen in dialysis-associated amyloidosis.

Question 3: This patient is most likely to also have which of the following conditions?

A. Systemic amyloidosis

B. Sjögren syndrome

C. Calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, and telangiectasia syndrome

D. Underlying malignancy

E. Primary biliary cirrhosis

Answers:

A. Incorrect - Although the occurrence of systemic amyloidosis in patients with nodular cutaneous amyloidosis was once thought to be as high as 50%, more recent studies have shown that nodular cutaneous amyloidosis only poses a 4%-15% chance of progressing to systemic amyloidosis.[2]

B. Correct - Studies have shown that as many as 22% of patients with nodular cutaneous amyloidosis are also diagnosed with Sjögren syndrome.[4]

C. Incorrect - Although nodular amyloidosis is associated with calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, and telangiectasia syndrome, Sjögren syndrome has demonstrated a much stronger association.[4,5]

D. Incorrect - Although nodular cutaneous amyloidosis can be associated with systemic amyloidosis, which is seen in patients with malignancies such as multiple myeloma, Sjögren syndrome is far more common in these patients.[2]

E. Incorrect - Primary biliary cirrhosis is weakly associated with nodular cutaneous amyloidosis but not to the same degree as Sjögren syndrome has demonstrated.[2]

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