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Case Presentation

Primary amyloidosis-induced nail dystrophy

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Abstract

Primary amyloidosis is caused by a monoclonal proliferation of plasma cells and is capable of producing cutaneous lesions. A 56-year-old male was admitted to the hospital for evaluation of chronic back pain and acute lower extremity weakness. On examination, he was noted to have subungual verrucous plaques with overlying nail dystrophy on his bilateral thumbs. A biopsy of one subungual lesion showed a deposition of amorphous material in the dermis that stained with Congo red and crystal violet. These cutaneous lesions ultimately led to the diagnosis of plasma cell dyscrasia and primary amyloidosis. Based on the literature search, primary amyloidosis presenting with this degree of subungual thickening and overlying nail dystrophy has not been previously reported.

Keywords: Primary amyloidosis, Nail dystrophy, Plasma cell dyscrasia

Introduction

Primary amyloidosis is caused by a monoclonal proliferation of plasma cells and produces cutaneous lesions in up to 40% of patients [1-4]. The lesions typically seen are waxy, purpuric papules often located in the flexural, facial, anogenital, and oral regions [5,-7]. We present an unusual case in which the initial manifestation of primary amyloidosis was subungual verrucous plaques with overlying nail dystrophy.

Case Report

A 56-year old-man with past medical history of hypertension, chronic back pain, and bilateral carpal tunnel syndrome was admitted for evaluation of worsening back pain of several months duration and acute lower extremity weakness. A dermatology consult was placed for evaluation of “warts” on his thumbs bilaterally. On physical exam, the patient had significant subungual verrucous plaques with overlying nail dystrophy on his bilateral thumbs (Figure 1). The patient reported that the lesions had been present for 2 years. He claimed that they were painful and continuously increasing in size. The patient also had waxy flesh-colored papules at his hairline (Figure 2a) and infiltrative plaques in his bilateral conchal bowls (Figure 2b).



Figure 1. Subungual verrucous plaques with overlying nail dystrophy on the patient's left (a) and right (b) thumb.



Figure 2. a-Waxy, flesh-colored papules at the patient's hairline. b-Infiltrative, waxy plaques in the patient's conchal bowl.

Significant laboratory examinations included a normocytic anemia, elevated prothrombin time, elevated ESR, elevated CRP, elevated BUN, bilirubinemia, decreased albumin, elevated alkaline phosphatase, elevated ALT, and decreased serum immunoglobulins (IgG, IgA, IgM). There was an M spike present in the beta-gamma region on plasma electrophoresis. Urine was positive for an elevated Kappa-to-Lambda light chain ratio and Bence Jones protein. Blood cultures were positive for MRSA. Serological assays were negative for HIV 1 and 2 antibodies, Hepatitis B antigen, and Hepatitis B antibody. MRI showed multiple compression fractures of the spine, increased gallium uptake around the left proximal femur, and opacities around the humeral head consistent with osteochondromatosis. Pathologic examination of the skin lesions revealed a dermal deposition of amorphous eosinophilic material with prominent clefts and sparse to absent inflammation (Figure 3a). The eosinophilic material stained strongly with Congo red (Figure 3b) and crystal violet. A plasma cell dyscrasia of 8-9% of kappa monoclonal plasma cells was detected in the bone marrow via aspiration and biopsy.

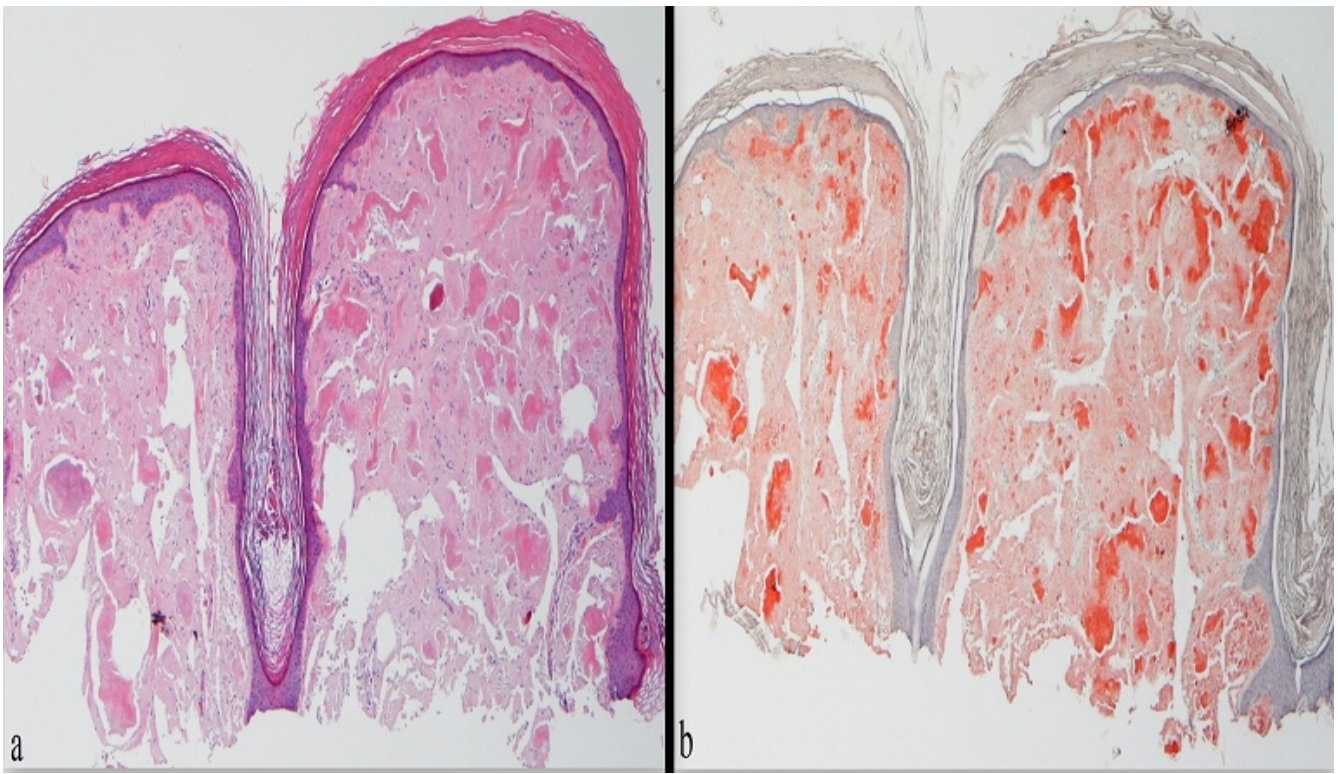


Figure 3: a-H&E of the thumb lesion showing dermal deposition of eosinophilic material. b-Congo red stain of the thumb lesion.

The patient was diagnosed with primary amyloidosis and was treated with bortezomib and dexamethasone. The patient's bacterial infection was successfully treated with intravenous vancomycin and piperacillin/tazobactam (Zosyn). He is currently being evaluated for bone marrow transplantation.

Discussion

Primary amyloidosis is defined as a clonal proliferation of plasma cells that produces excess amounts of immunoglobulin light chains (also known as AL proteins), which deposit into extracellular tissues [1-2]. Cutaneous lesions are seen in up to 40% of patients with primary amyloidosis owing to amyloid deposition in the dermis [3-4]. Histologically, these deposits stain with crystal violet and will have an apple-green birefringence when stained with Congo red and viewed with a polarized light source. Typically, the cutaneous lesions of primary amyloidosis present as waxy, purpuric lesions, often located in the flexures, eyelids, anogenital region, and oral cavity [5-7]. Less common cutaneous manifestations are infiltrative lesions, vesicles, chronic paronychia, and nail dystrophy [4,6-11].

The changes seen in primary amyloidosis-induced nail dystrophy have been previously described as similar to those seen in lichen planus: brittleness, longitudinal ridging, onychoschizia, subungual thickening, onycholysis, and anonychia [4,6,12-13]. There have not been any cases of subungual verrucous plaques with overlying nail dystrophy to this degree previously reported.

Subungual verrucous plaques with overlying nail dystrophy is a rare initial presentation of primary amyloidosis. The differential diagnosis would include fibrokeratoma, Koenen tumor, verrucae, verrucous carcinoma, and squamous cell carcinoma. Increased suspicion for primary amyloidosis is necessary if patients with dystrophic nails or subungual verrucous plaques have clinical signs and symptoms indicative of the disease such as macroglossia, weight loss, fatigue, carpal tunnel syndrome, or hepatosplenomegaly.

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