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CASE REPORT

CUTANEOUS AMYLOIDOSIS IN A PATIENT WITH SYSTEMIC AMYLOIDOSIS DUE TO MULTIPLE MYELOMA

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Abstract

Cutaneous manifestations in systemic amyloidosis secondary to multiple myeloma or AL amyloidosis are seen in approximately 15%-40% of patients with systemic amyloidosis. Cutaneous involvement has a preference for skin folds, retroauricular region, eyelids, neck and the axilla and may present in the form of purpura, domed papules or nodes resembling isolated nodular amyloidosis. Congo red staining remains the gold standard for diagnosis, revealing amyloid fibrils with apple-green birefringence in the polarized microscopy. It is noteworthy that patients may sometimes present with non-specific skin changes, suggestive of depositional disease, such as alopecia or nail dystrophy. Herein, we present a patient with systemic amyloidosis, who exhibited red-brown macules coalescing into a rippled pattern in the extremities, amyloid nodules in the peri-auricular region and dystrophic nails, findings that were attricuted to cutaneous amyloidosis in the context of systemic amyloidosis.

Keywords: Amyloidosis, systemic, cutaneous, erythema, dystrophic nails.

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CASE STUDY

A sixty-nine years old male patient presented to the hospital with diarrhea and a rash, mainly on the face and hands, which appeared almost simultaneously three months ago. The rash was erythematosous and pruritic with mild scaling, while nail dystrophy was noted, too (Figure 1). From his past medical history, the patient was diagnosed three years ago with multiple myeloma, as the bone marrow biopsy then showed infiltration with plasma cells of 45% and he was administered bortezomib, cyclophosphamide and dexamethazone. However, he subsequently commenced haemodialysis for the last year and he was only on lenalinomide treatment thereafter.

From the laboratory tests, the abnormal findings were the following: Ht: 30%, Hb: 9.1g/dL, WBC:3.750 cells/m^3 (45% neutrophils, 44.3% lymphocytes), PLTs: 88.000 cells/μL, hs-CRP: 17,7 mg/dL (normal range <0.5 mg/dL), serum potassium levels: 2.9 mmol/L, (normal range: 3.5-5 mmol/L), urea: 119 mg/dL (normal range <45mg/dL), serum creatinine: 6.5 mg/dL (normal range <1.5 mg/dL). Stool testing for *Clostridioides difficile* toxin was negative thrice, as were three consecutive stool cultures.

The patient underwent a skin biopsy, the histopathology of which revealed the deposition of eosinophilic material in the dermis, that tested positive in Congo red. In addition, due to the multiple diarrheas, we performed a sigmoid colon biopsy, which tested positive in Congo red, too. A diagnosis of systemic amyloidosis due to multiple myeloma was confirmed and the patient was transferred to the Hematology Department for further management. Notably, before further treatment, a heart ultrasound was performed, which revealed an ejection fraction of 35%-40% with diffuse hypokinesias.

Cutaneous manifestations in systemic amyloidosis secondary to multiple myeloma or AL amyloidosis are seen in approximately 15%-40% of patients with systemic amyloidosis.[1,2] Cutaneous involvement has a preference for skin folds, retroauricular region, eyelids, neck and the axilla and may present in the form of purpura, domed papules or nodes resembling isolated nodular amyloidosis.[3,4] Biopsy is essential for confirming the diagnosis and should be performed. Histopathologic findings include deposits of an eosinophilic, amorphous material, called amyloid deposit, with the eosin and hematoxylin staining in the dermis, which may sometimes invade the walls of dermal blood vessels. Congo red staining remains the gold standard for diagnosis, revealing amyloid fibrils with apple-green birefringence in the polarized microscopy.[5] It is noteworthy that patients may sometimes present with non-specific skin changes, suggestive of depositional disease, such as alopecia or nail dystrophy.[6,7] Our patient exhibited red-brown macules coalescing into a rippled pattern in the extremities, amyloid nodules in the peri-auricular region and dystrophic nails. The highly heterogeneous clinical picture, the difficulty in diagnosing systemic amyloidosis due to its heterogeneity as well as difficulties in biopsying the right infiltrated organ, together with the high mortality rate of the disease, highlight the importance of the cooperation of many clinicians as well as pathologists for the accurate diagnosis of this perplexed clinical entity.

There is no conflict of interest regarding this manuscript.

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ANNEX

Figure 1. Red-brown macules coalescing into a rippled pattern on the anterior surface of the shin.