A case of plasma cell dyscrasia presenting as nonscarring alopecia

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INTRODUCTION
Amyloidosis is a term often used to describe diseases in which a fibrillar proteinaceous material, amyloid, is deposited in the vasculature and tissue of different organs and can present with a wide variety of manifestations. Amyloid itself is a breakdown product of various proteins whose origins include, but are not limited to, the skin, blood, brain, and liver. Although it has many sources, all variants share a common configuration molecularly, that is, the cross-β pleated sheet. On histopathology, deposits of pink amorphous material can be seen with hematoxylin-eosin stain, which characteristically are birefringent when stained with Congo red and viewed under polarized light.1

Amyloid can be deposited locally in the skin in the form of localized cutaneous amyloidosis or as a manifestation of a systemic process such as a plasma cell dyscrasia or chronic inflammatory states (eg, rheumatoid arthritis). Localized and systemic cutaneous amyloidosis can be further subdivided into primary and secondary, each with its unique set of underlying causes.

We recently encountered a patient who presented with diffuse alopecia on her scalp that was subsequently biopsied, revealing findings consistent with amyloidosis. Subsequent workup diagnosed an underlying systemic process.

CASE REPORT
A 65-year-old Black woman with a past medical history of hypertension, hyperlipidemia, and diabetes mellitus presented for diffuse hair loss over a 6-month period. Her hair loss was associated with periods of intense itching, burning, and tenderness. No prior treatments had been trialed, and she denied fever, chills, rash, joint pain, or joint stiffness.

Physical examination revealed diffuse scaling without erythema and patchy areas of alopecia involving the majority of her scalp, sparing only the mid-lower portion of the occipital scalp (Fig 1). Hair pull test was negative, and no specific findings were seen on dermoscopy. Although the scaling was attributed to seborrheic dermatitis, the distribution of the alopecia was peculiar given her history, and thus a punch biopsy was performed. The biopsy revealed deposition of eosinophilic material within the dermis, around hair follicles, and focally within some small subcutaneous vessels (Fig 2). The deposits were positively stained with Congo red and were birefringent under polarized light. As these histologic findings were consistent with a diagnosis of amyloidosis, an underlying systemic workup was initiated.

Complete blood cell count, complete metabolic panel, and both serum and urine protein electrophoresis were ordered, and the patient was referred to medical oncology for further management. The complete blood cell count and metabolic panel revealed no pertinent abnormalities. Serum and urine protein electrophoresis revealed no M spike; however, her serum kappa-to-lambda ratio was 0.06. While the patient was awaiting her oncology appointment, she developed lower extremity edema, and she was referred to cardiology, whose workup did not reveal an underlying cardiac etiology. She was then sent to nephrology, and a workup at that time showed an elevated creatinine level of 30 mg/dL.

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1.24 mg/dL, total protein of 3.8 g/dL, and albumin of 2.0 g/dL. The oncology team performed a bone marrow biopsy that revealed signs concerning for a plasma cell dyscrasia. She was started on bortezomib, dexamethasone, and cyclophosphamide and was responding to therapy appropriately, without any improvements in her alopecia, with plans for autologous stem cell transplantation in 2021.

**DISCUSSION**

Systemic amyloidosis can affect any organ system and classically can lead to nephrotic syndrome, restrictive cardiomyopathy, peripheral neuropathy, hepatomegaly, macroglossia, purpura, and bleeding diathesis. Although alopecia is an uncommon finding seen in systemic amyloidosis, it has been described in the literature. Lutz et al\(^2\) described a case of generalized alopecia secondary to systemic amyloidosis that presented 6 years prior to any other systemic manifestation. Additionally, Magro et al\(^3\) described a case series of 3 patients with nonscarring diffuse alopecia. In their report, 1 patient had localized amyloidosis, whereas the other 2 had plasma cell dyscrasias. The reason for alopecia occurrence in these patients has not been fully elucidated; however, Hunt et al\(^4\) suggested that

![Fig 1. Patchy alopecia involving the majority of the scalp but sparing the mid-lower portion of the occipital scalp.](image)

![Fig 2. A. Perifollicular amyloid deposition. B. Sebaceous amyloid deposition. C. Vascular amyloid deposition. D. Amyloid deposition in adipose tissue. (A, B, C, and D, Hematoxylin-eosin stain; original magnifications: A, ×100; B, ×100; C, ×100; D, ×100.)](image)
amyloid deposition within hair follicles prematurely transitions the hair cycle into catagen. These catagen hairs then progress to “normal” telogen hairs and persist in this stage, explaining the progressive and prolonged alopecia in untreated patients. Another theory, which they believed was less likely, involved defective anagen maturation secondary to compression of the hair follicle sheath.

We highlight yet another case of diffuse alopecia secondary to amyloid deposition to raise awareness of this uncommon presentation of amyloidosis and plasma cell dyscrasias. Scalp biopsies are an invaluable tool for all forms of hair loss and should be utilized in perplexing cases. In this scenario, a diagnosis of amyloidosis warrants systemic evaluation for an associated underlying disorder. Initial laboratory workup should include a complete blood cell count, complete metabolic panel, and both serum and urine protein electrophoresis for evaluating monoclonal gammopathy.1 Systemic amyloidosis can be a devastating disease. Mortality in late-stage disease has been estimated to be approximately 4 to 6 months. Thus, clinicians should be vigilant in presentations that may fit its many manifestations, as an early treatment can greatly impact mortality.

Conflicts of interest
None disclosed.

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