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An Elderly Male With Progressive Nail Atrophy: Answer

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An Elderly Male With Progressive Nail Atrophy: Answer

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ANSWER

Systemic light-chain amyloidosis (AL).

DISCUSSION

Histopathological examination of the nail bed revealed a pink amorphous material surrounding blood vessels in the dermis (Figure 2 in the Question portion). A Congo red stain highlighted the material (Fig. 3), which also demonstrated faint green-yellow birefringence on polarization (not shown). Immunohistochemistry for lambda light chains was positive (Fig. 4), whereas there was no staining for kappa. A previously obtained gastric biopsy from 6 months before was revisited and was also found to demonstrate amyloid on Congo red staining.

Given these pathologic findings combined with the patient's constellation of symptoms, a diagnosis of immunoglobulin light-chain (AL) amyloidosis was strongly suspected and a thorough systemic workup was recommended. A serum protein electrophoresis study was obtained and revealed a small free lambda monoclonal protein (<0.1 g/dL) with marked suppression of polyclonal gammaglobulin. A free serum light chain analysis revealed a lambda level of 2686.0 (normal range: 5.7–26.3 mg/L). A subsequent bone marrow biopsy revealed atypical plasma cells occupying 25% of the marrow space consistent with plasma cell myeloma.

The patient was treated with 3 cycles of bortezomib, cyclophosphamide, and dexamethasone (CyBorD) with poor response and disease progression. He was not eligible for stem cell transplant due to severe amyloid-included restrictive cardiomyopathy complicated by worsening congestive heart failure. Three months after his diagnosis, he was admitted to hospice for comfort measures.

AL amyloidosis is a paraneoplastic phenomenon characterized by deposition of misfolded proteins in various organs. The most commonly affected organs include the heart, kidneys, liver, soft tissues, peripheral and/or autonomic nervous system, and gastrointestinal tract.[1,2] It is associated with an underlying plasma cell dyscrasia, usually multiple

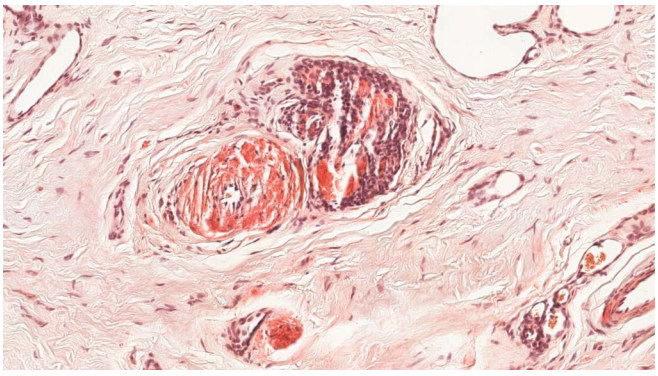

FIGURE 3. Congo red stain (original magnification, ×400).

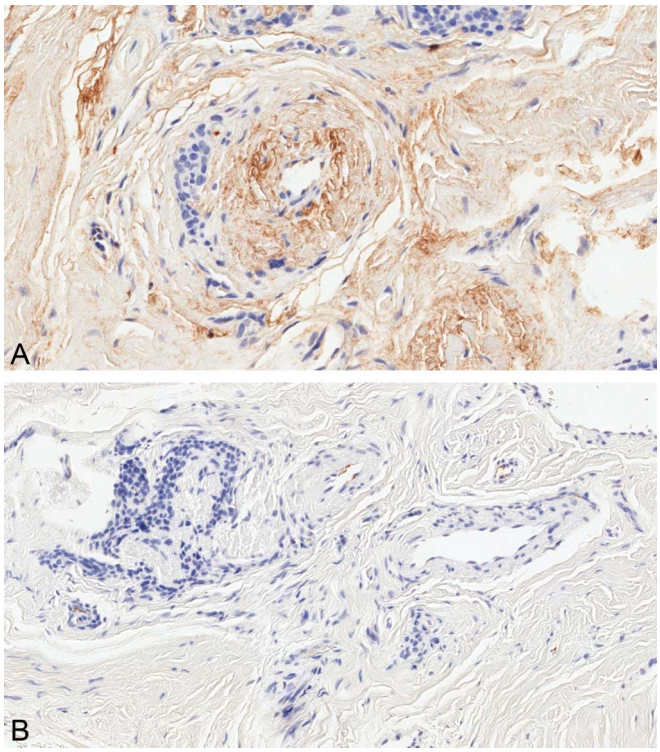

FIGURE 4. A, Lambda immunohistochemical stain (original magnification, ×400). B, Kappa immunohistochemical stain (original magnification, ×400).

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myeloma. Often, the myeloma is diagnosed subsequent to the amyloid-induced systemic complications, as seen in our case. Presenting symptoms of systemic AL amyloidosis are broad and may mimic other more common diseases seen in elderly patients and unfortunately this can promote diagnostic delay.

Mucocutaneous involvement in systemic AL amyloidosis occurs in one-third of patients including those with the myeloma-associated form and is rarely the initial manifestation.[2–5] Commonly reported findings are periorbital "pinch" purpura and macroglossia. Sporadic cases of AL amyloidosis presenting as 20 nail dystrophy have also been reported.[3–5] Dermatologists and dermatopathologists must be aware of this association because they may be relied upon to make an earlier diagnosis. Optimal treatment is controversial with poor outcomes overall. High-dose steroids, chemotherapeutic regimens including bortezomib-based regimens or melphalan, and stem cell transplantation are among the modalities typically used.[6–8]

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