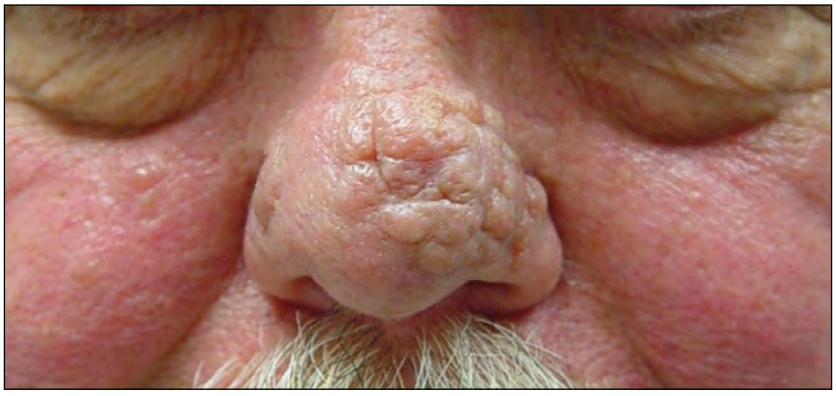
What Is Your Diagnosis?



A 66-year-old man presented with lesions growing on the nose of 5 years' duration. He reported intermittent bleeding and purulent discharge from the lesions but denied any associated pain or pruritus. He was treated for rosacea, alternating between tetracycline and doxycycline for 4 years prior to presentation. Despite treatment, the lesions steadily increased in number and size. The patient's medical history was otherwise unremarkable, and he lacked any constitutional symptoms.

PLEASE TURN TO PAGE 283 FOR DISCUSSION

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The authors report no conflict of interest.

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The Diagnosis: Primary Localized Cutaneous Nodular Amyloidosis

The patient presented with multiple waxy, pink and flesh-colored papules and nodules on the nasal bulb, predominantly on the left side, with involvement of the nasal ala (Figure 1). The lesions were soft and nontender. Serum protein electrophoresis and immunofixation to investigate systemic involvement revealed a normal pattern. The patient's complete blood cell count was within reference range and an antinuclear antibody test was negative. A shave biopsy from the left side of the nose was performed. The hematoxylin and eosin–stained sections revealed diffuse dermal globular and perivascular eosinophilic deposits with destruction of normal collagen and elastic fibers (Figure 2). Congo red stain confirmed amyloid deposition and showed apple green birefringence when viewed under polarized light (Figure 3). The lesions were removed via shave excision, followed by electrodesiccation and curettage. The patient showed no signs of recurrence at 18-month follow-up (Figure 4).

Amyloidosis is a group of diseases characterized by extracellular deposits of insoluble amyloid fibrils.[1] There are at least 18 unrelated precursor proteins to amyloid, resulting in different clinical presentations.[2] The 2 major clinical classifications are localized and systemic.[3] Cutaneous amyloidoses are localized forms involving amyloid deposits exclusively in the dermis. Primary localized cutaneous nodular amyloidosis (PLCNA), the rarest of the primary cutaneous amyloidoses,[4] presents with deposits of AL protein throughout the dermis, in contrast to macular and lichen amyloidosis, which have keratin-derived AK protein deposits solely in the papillary dermis.[5] Immunohistochemical evidence suggests that the AL protein originates from immunoglobulin light chains secreted by local plasma cells.[6] Systemic amyloidosis may present with histologically identical AL protein deposits, but deposition in multiple vital organs also will occur.

Primary localized cutaneous nodular amyloidosis presents as single or multiple pink to brown nodules that most commonly appear on the legs, followed by the head, trunk, arms, and genitalia, respectively.[7] Clinical morphology of lesions may be atrophic, anetodermic, or bullous, with differences likely attributable to collagen and elastic fiber destruction.[8] Rare cases of lesions presenting as plaques rather than

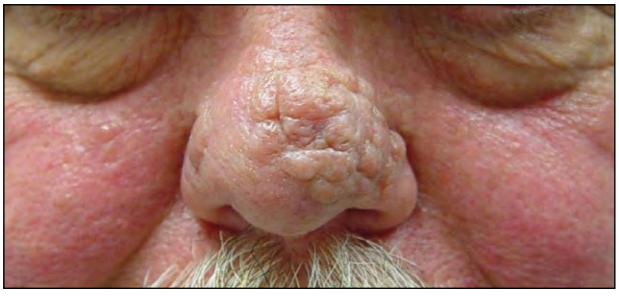

Figure 1. Numerous pink and flesh-colored papules and nodules on the nose.

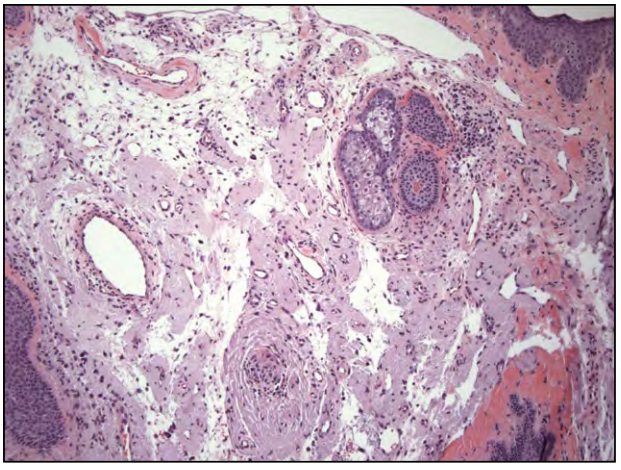

Figure 2. Diffuse dermal globular and perivascular eosinophilic deposits (H&E, original magnification ×100).

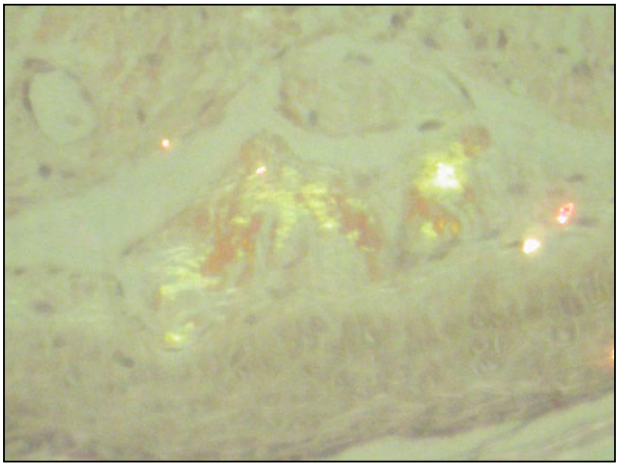

Figure 3. Apple green birefringence of amyloid deposits under polarized light (Congo red, original magnification ×200).

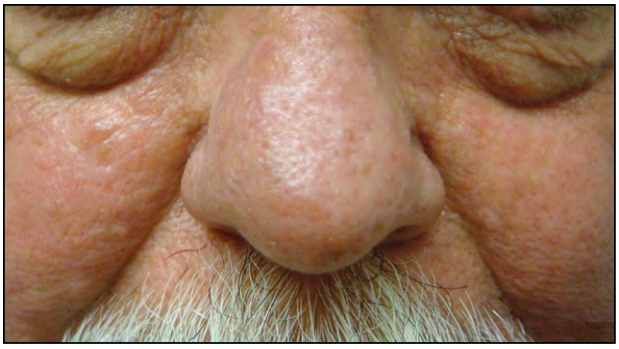

Figure 4. Absence of recurrent lesions at 18-month follow-up.

nodules have been reported.[5,8] Disease onset predominantly occurs between the 40th and 60th decades of life, with equal frequency in men and women.[9]

Although PLCNA runs a benign course and treatment largely is focused on cosmetic or functional concerns, systemic amyloidosis carries a much poorer prognosis. Therefore, on diagnosis of PLCNA, it is essential to rule out systemic amyloidosis or underlying plasma cell dyscrasia.[10] The suggested rate of progression of the cutaneous form to systemic disease varies. The rate may be much lower now than previously had been reported (7% vs 50%)[10]; however, this lower rate is still being debated.[11] Multiorgan involvement may be evaluated using serum amyloid P component scintigraphy, a scanning technique using radiolabeled serum amyloid P component to reveal amyloid deposits in vivo.[10] Primary localized cutaneous nodular amyloidosis has been reported in association with several other connective-tissue diseases, such as Sjögren syndrome and CREST (calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, and telangiectasia) syndrome.[12,13]

Various treatments have been recommended for PLCNA, including cryotherapy; dermabrasion[4]; electrodesiccation and curettage; and surgical excision, with and without split-thickness grafting.[14] Intralesional steroid injection generally is ineffective and may even increase amyloid deposition.[15] Newer therapeutic approaches using pulsed dye and CO_2 lasers reportedly delivered success but not without complications.[3,16,17] Unfortunately, none of the aforementioned therapeutic approaches have been proven to be consistently effective, and recurrence of the nodular lesions is common.[10]

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