Nodular Amyloidosis: Case Report and Literature Review

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Abstract

Background: Amyloidosis refers to a group of depositional diseases that are classified into two main types: systemic and localized. Large nodules of localized cutaneous amyloidosis of the nasal ala and surrounding skin are rare and the treatment is often unsatisfactory.

Objective: We report a case of rapidly enlarging, localized, nodular cutaneous amyloidosis of the nose and the surrounding skin with a brief review of the current literature regarding treatment of this rare disease.

Conclusion: Nodular amyloidosis can be treated successfully with cold steel excision in combination with carbon dioxide laser. Close followup of these patients is warranted, as nodular amyloidosis may be the precursor to systemic amyloidosis.

We report a case of rapidly enlarging, localized, nodular cutaneous amyloidosis with a review of the literature pertaining to this rare form of amyloidosis and its treatment.

Case Report

A wheelchair-bound 67-year-old male nursing home resident was referred to the Department of Otolaryngology-Head and Neck Surgery for evaluation and treatment of large growths on his nose and the surrounding skin. The nasal masses had enlarged progressively for two to three years and were now causing difficulty with both breathing and eating. The masses were so large that they had to be lifted in order to feed the patient (Fig. 1). His past medical history was significant for diabetes, epilepsy, schizophrenia, organic brain syndrome, and cerebrovascular accident. Cutaneous examination revealed two shiny,
yellow to reddish brown nodules, measuring 6 x 5 x 3 cm on the left nasal ala and 6 x 5.5 x 4.5 cm on the right nasal ala. In addition there were smaller papules on the upper lip and cheek extending up to 4 cm from the nose laterally on each side. The lesions were not painful or pruritic. A differential diagnosis of nodular rhinophyma, which is much more common in this region, was considered.

Results of (Complete Blood Count), platelet count, serum protein electrophoresis, and immunoelectrophoresis for M protein were within normal limits. Antinuclear antibody was negative. Alkaline phosphatase was slightly elevated (161 U/L) as was the sedimentation rate (48 mm/h).

Surgery was carried out under general anesthesia with an orotracheal tube. The large nodules were first excised using a No. 15 blade. The area was then sculpted, and hemostasis was achieved with a carbon dioxide laser. Thus, a near-total excision of the nodules was carried out. After this procedure the patient was able to breathe through his nose very well.

The histologic observation of the paraffin-embedded tissue revealed a nodule covered by an atrophic epidermis with a few sebaceous glands seen in the dermis. Stromal fibrosis was prominent, and an infiltration with lymphocytes and perivascular plasma cells was noted. Extensive deposits of pink amorphous material were noted in both nodules, filling almost the entire dermis; this material stained positive with Congo red (Fig. 2). When viewed under a polarized light, the material stained with Congo red exhibited a green birefringence indicating an amyloid deposition.

Immunohistochemical studies were carried out on paraffin-embedded tissue using anti-kappa and anti-lambda light chain antibodies only and revealed equivocal positive results for both kappa and lambda light chains, which constitute the AL amyloid. The monoclonality of the infiltrating plasma cells was not specifically evaluated. No signs of amyloid formation have been seen at any other body sites over the past year. Based on the results of the clinical and histologic examination, nodular cutaneous amyloidosis was diagnosed.

Discussion

Amyloidosis refers to a family of disorders in which an abnormal extracellular deposition of protein occurs causing alterations of tissue architecture and function.1 Amyloid deposits, regardless of clinicopathologic type or tissue involvement, are composed of paired 7.5-10-nm straight, rigid, nonbranching, hollow fibrils arranged in a loose meshwork.2 These fibrils undergo a degenerative process which transforms the natural alpha-helical structure into the classic antiparallel, beta-pleated sheet characteristic of amyloid.3,4 It is this configuration that probably accounts for the fibrils' ability to bind the Congo red stain. The deposits demonstrate an apple-green birefringence when viewed under polarized light with Congo red stain, crystal violet, and thioflavine T fluorescence.5

Amyloidosis is classified into two types based on its distribution: systemic or localized. These groups are further divided according to the type of amyloid fibril protein (amyloid A, amyloid L, or keratin), clinical appearance, and the pathogenic mechanism of deposition.

The systemic group includes primary amyloidosis, which is due to an underlying plasma cell dyscrasia, myeloma-associated amyloidosis, and secondary systemic amyloidosis. Causes of the latter include chronic infection, rheumatoid arthritis, inflammatory bowel disease, Hodgkin's disease, solid nonlymphoma tumors, Bechet's syndrome, Reiter's syndrome, and Sjogren's syndrome.6,7