

Single Case

Nodular Cutaneous Amyloidosis at the Temple

Kathrin Schucht^a Josef Schröder^b Heiko Siegmund^b Claudia Grafe^b
Stephan Schreml^a

^aDepartment of Dermatology, University Medical Center Regensburg, Regensburg, Germany; ^bInstitute of Pathology, University of Regensburg, Regensburg, Germany

Keywords

Amyloid · Amyloidosis · Primary localized cutaneous nodular amyloidosis

Abstract

A 52-year-old woman presented with a large partially yellow and erythematous tumor on her right temple. She reported that it had grown over the last 4 years. Regional lymph nodes were impalpable. A punch biopsy showed eosinophilic material in the dermis and subcutis. Immunohistochemistry showed positive staining for kappa and lambda light chains. Electron microscopy showed the typical amyloid fibrils (7–10 nm in diameter). There was no evidence of systemic amyloidosis, paraproteinemia or underlying plasmacytoma. The tumor was completely removed via curettage. At follow-up, the patient presented in good health with no signs of relapse.

© 2016 The Author(s)
Published by S. Karger AG, Basel

Introduction

Amyloidoses originate from the extracellular aggregation of autologous protein fibrils. Within these aggregations, the pentagonal amyloid P component serves as a stabilizer molecule. Amyloidoses can be classified into three categories: primary localized cutaneous amyloidosis (PLCA), secondary localized cutaneous amyloidosis, and systemic amyloidosis with cutaneous involvement [1]. In the PLCA group, three types can be distinguished: macular,

popular (lichenoid) and nodular forms [1–4]. The first two are mainly located on the trunk, and cytokeratins serve as the amyloid precursors. Here, amyloid deposition is limited to the papillary dermis [1–5]. However, in the rare cases of nodular PLCA, amyloid consists of aggregated kappa and lambda light chains, which can be found both in the dermis and the subcutaneous tissue [1–4, 6]. Furthermore, nodular PLCA can originate from systemic amyloidosis or progress to systemic disease [3, 4]. Here, we present a case of nodular PLCA on the temple of a 52-year-old woman.

Case Report

A 52-year-old lady presented with a soft, shiny, partially yellow, erythematous tumor (3.5 × 4.5 cm) with telangiectasia on her right temple (fig. 1a, b). She recalled having had this tumor for about 4 years and that it had significantly grown lately. She negated pruritus. Regional lymph nodes were impalpable. A punch biopsy showed amorphous eosinophilic material within the dermis and subcutis (fig. 1c, d) which was positive in Congo red staining. In immunohistochemistry, the stainings for kappa and lambda light chains were positive (fig. 1e, f). The diagnosis was confirmed by electron microscopy, and the typical amyloid fibrils (7–10 nm in diameter) were found (fig. 1g, h) [3]. The tumor was completely removed via curettage. Additionally, a punch biopsy from abdominal fat tissue was examined via electron microscopy [7]. There was no evidence of systemic amyloidosis. An electrocardiogram and abdominal sonography showed no pathologies. There were no signs of paraproteinemia or underlying plasmacytoma. Therefore, the diagnosis was nodular PLCA. A punch biopsy taken during follow-up showed some remaining amyloid so that the patient is now scheduled to undergo a second surgery via curettage. Regular check-ups are scheduled to detect a potential progress to systemic disease.

Discussion

The diagnosis of PLCA requires histological analysis of a skin specimen, complemented by immunohistochemistry and electron microscopy. Histologically, eosinophilia as well as positivity in periodic acid-Schiff, Congo red, and thioflavin T stainings are characteristics of amyloid deposits [1, 8]. Immunohistochemistry with antibodies directed against cytokeratin and immunoglobulin light chains (lambda and kappa) allows to further distinguish between the different forms of PLCA. Electron microscopy confirms the diagnosis of amyloidosis when the typical amyloid fibrils (7–10 nm in diameter) are found. Image quality improves significantly when the skin sample is fixed in Karnofsky's fixative (glutaraldehyde) instead of paraformaldehyde [4]. For patients with nodular PLCA, it is recommended to assess for progression to systemic amyloidosis on a regular basis. This should include a full history and physical examination along with electrocardiogram, complete blood count, serum creatinine levels, serum liver-associated enzyme levels, serum electrophoresis, and urine examination [1]. Progression to systemic disease has been described to occur in 5–50% of cases [1]. Furthermore, an abdominal fat biopsy has been suggested as an easy method to detect a potential progression to systemic disease [3, 7]. Nodular PLCA is extremely rare, accounting for around 1.5% of PLCA cases [1]. It has to be mentioned though that nodular PLCA is more frequent in Asia and South America when compared to Europe or North America.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors declare no conflicts of interest.

References

- 1 Schreml S: Cutaneous amyloidoses; in Griffiths C, Barker J, Bleiker T, et al (eds): Rook's Textbook of Dermatology, ed 9. Chichester, John Wiley & Sons, 2016, pp 58.1–58.13.
- 2 Breathnach SM: Amyloid and amyloidosis. *J Am Acad Dermatol* 1988;18:1–16.
- 3 Schreml S, Szeimies RM, Vogt T, Landthaler M, Schroeder J, Babilas P: Cutaneous amyloidoses and systemic amyloidoses with cutaneous involvement. *Eur J Dermatol* 2010;20:152–160.
- 4 Haverkamp S, Evert K, Schröder J, Schreml S: Nodular cutaneous amyloidosis resembling a giant tumor. *Case Rep Dermatol* 2016;8:22–25.
- 5 Brownstein MH, Helwig EB: The cutaneous amyloidoses. I. Localized forms. *Arch Dermatol* 1970;102:8–19.
- 6 Ritchie SA, Beachkofsky T, Schreml S, Gaspari A, Hivnor CM: Primary localized cutaneous nodular amyloidosis of the feet: a case report and review of the literature. *Cutis* 2014;93:89–94.
- 7 Westermark P, Davey E, Lindbom K, Enqvist S: Subcutaneous fat tissue for diagnosis and studies of systemic amyloidosis. *Acta Histochem* 2006;108:209–213.
- 8 Li WM: Histopathology of primary cutaneous amyloidoses and systemic amyloidosis. *Clin Dermatol* 1990;8:5–30.

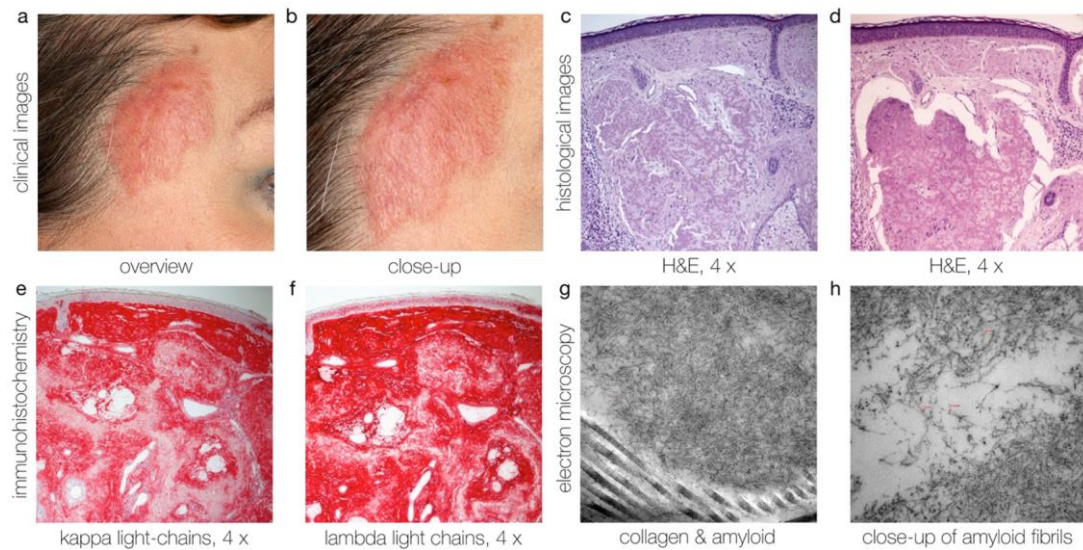


Fig. 1. **a, b** Large tumor on the right temple of a 52-year-old woman. The sharply demarcated tumor exhibited a shiny, yellow-reddish surface with some telangiectasias. **c, d** A punch biopsy showed amorphous eosinophilic material in the dermis and subcutis. **e, f** Immunohistochemistry revealed massive amounts of both kappa and lambda immunoglobulin light chains. **g, h** Under electron microscopy, the typical amyloid fibrils (7–10 nm in diameter) were seen.