

## Single Case

# A Peculiar Case of Large and “Unresectable” Primary Localized Cutaneous Nodular Amyloidosis of the Ankle

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## Keywords

Cutaneous amyloidosis · Nodular amyloidosis · Surgical treatment

## Abstract

Nodular cutaneous amyloidosis represents the rarest variant of primary localized cutaneous amyloidosis. The proposed management ranges from topical or systemic agents to surgical treatment. Complete surgical excision is advisable due to its potential progression to systemic amyloidosis due to dermis and subcutaneous tissue infiltration. However, in particular locations, the risk of functional complications is high, so an alternative treatment option should be considered. We report a case of a large primary nodular cutaneous amyloidosis of the leg involving the joint capsule which was successfully treated by incomplete surgical removal, without recurrences at 7-year follow-up.

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## Introduction

Primary localized cutaneous amyloidosis (PLCA) is characterized by extracellular deposition of amyloid proteins in the skin without systemic involvement. Nodular cutaneous amyloidosis (NCA) is the rarest variant with <100 cases described, and its clinical presentation includes solitary or multiple lesions in acral areas, face, skull, or genitalia [1, 2]. We

describe the case of a large primary NCA of the leg involving the joint capsule, treated by incomplete surgical removal, without recurrences at 7-year follow-up.

### Case Presentation

A 69-year-old male with no significant medical history presented with a cutaneous nodular lesion of the left leg that showed progressive growth in the last 9 months, resulting in patient functional limitations and difficulty in wearing standard shoes. Physical examination revealed a single, soft, bulky, cauliflower-like lesion on the medial malleolar region (Fig. 1a). Preoperative MRI showed a lesion measuring 10.8 × 9.9 × 4.7 cm with talocrural joint capsule involvement (Fig. 2); incisional biopsies confirmed a diagnosis of PLCA.

Therefore, a radical treatment would have included an en bloc removal of the lesion and joint capsule, resulting in joint exposure and ankle instability, ankle arthrodesis, and microsurgical reconstruction of the soft tissues defect. Conversely, the patient refused an invasive procedure and requested a debulking of the lesion in order to wear standard shoes and walk normally. A subtotal mass removal by shave excision and electrocoagulation with joint capsule preservation and subsequent reconstruction with skin grafts were performed. The histologic examination showed massive deposits of amorphous material in the dermal layer of the skin and subcutaneous tissue, with interstitial amyloid deposition and focal involvement of small blood vessels associated with an infiltrate of perivascular plasma cells stained with Congo red and Thioflavin (Fig. 3).

No evidence of systemic amyloidosis was found, and the patient refused any further radical treatment. At 7-year follow-up, no signs of recurrence or systemic amyloidosis were detected (Fig. 1b). The physiological function of the talocrural joint was preserved, and the patient was able to walk using standard shoes and to practice his favorite hobby and daily activities.

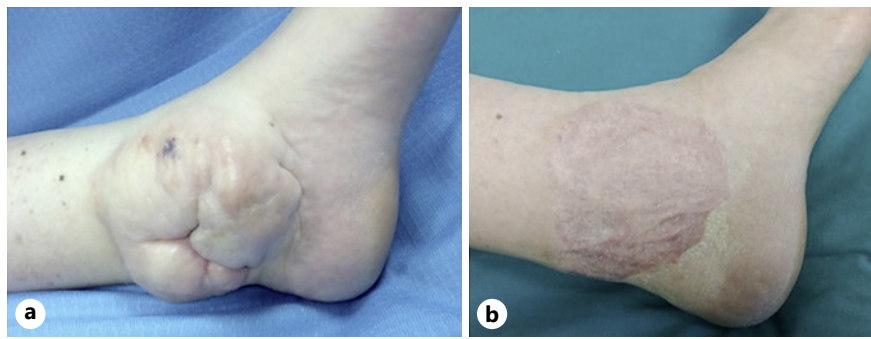
### Discussion

Cutaneous amyloidosis is rare and includes 3 clinical and histological subtypes: macular, lichenoid, and nodular. The peculiarity of the nodular variant consists in the potential progression to systemic amyloidosis due to dermis and subcutaneous tissue infiltration and the amyloid fibril composition. In fact, in NCA, the amyloid originates from immunoglobulin light chains (AL-type amyloid) and is associated with dermal plasma cell infiltration.

Several treatment strategies have been reported, encompassing topical and systemic agents, phototherapy, laser (CO<sub>2</sub> laser, ytterbium/erbium laser, pulsed dye laser, Erbium:YAG laser, and Nd:YAG laser), and surgical interventions such as excision, curettage, or shaving followed by dermabrasion or laser application; however, a recommendation on preferable procedures for PLCA is lacking. Surgical removal should be preferred for NCA because of the potential infiltration in subcutaneous tissue and blood vessels and progress to systemic amyloidosis [2, 3].

The present case implied a careful choice of the most appropriate treatment and represented a reconstructive challenge due to joint capsule infiltration. Basing on the benign nature of the lesion and the patient's consensus, we chose a more conservative treatment, thus avoiding functional limitations and insidious reconstructive procedures.

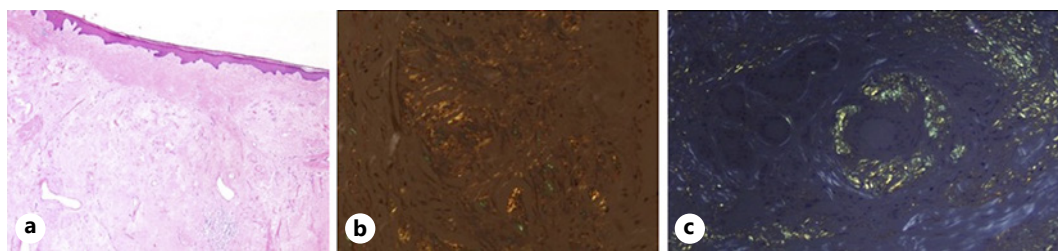
Despite the risk of local recurrences after surgical intervention due to the putative persistence of amyloids in the dermis, curettage or surgical shaving followed by laser application or cauterization has been proposed. Three reports reported the use of curettage for NCA of the head



**Fig. 1.** **a** Preoperative picture of the cutaneous lesion of the left leg. **b** 7-year follow-up picture showing no recurrence of the disease and an acceptable morphological result.



**Fig. 2.** Preoperative magnetic resonance image showing a lesion (red arrows) measuring 10.8 × 9.9 × 4.7 cm with talocrural joint capsule involvement.



**Fig. 3.** **a** Massive eosinophilic deposits in the dermis and hypodermis (hematoxylin and eosin. ×20). **b** Interstitial deposits of amyloid (Congo red stain under polarized light. ×100). **c** Perivascular green staining (Congo red stain under polarized light. ×100).

without recurrence [4–6]. Lien et al. [7] used a shave excision followed by dermabrasion without recurrence, as well as Grattan et al. [8] who reported 2 cases of NCA of the face.

We described a further case of NCA, and to the best of our knowledge, it is one of the largest and the first documented cases that showed involvement of a joint capsule. Although a complete eradication was not achieved, no local recurrence or progression to systemic amyloidosis was observed, supporting the literature.

An early dermatologic evaluation and diagnosis of these lesions remain essential because of the potential for infiltration, and the choice of the therapeutic strategy should be tailored to the clinical features of the lesion and patient's characteristics. While radical excision of NCA

is always recommendable, a conservative approach in critical areas which may require challenging surgical procedures should be considered.

### Statement of Ethics

The patient has given written informed consent to publish this case (including publication of images). Information revealing the patient's identity has been avoided. The research in this case report was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

### Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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### Author Contributions

A.S. performed the surgical procedure. M.T. performed analysis of the information and wrote the manuscript. F.B. performed the histological examination. B.P. and Y.Z. reviewed medical history of the patient and performed information search. G.D.B. reviewed the manuscript and supervised the project.

### Data Availability Statement

Further enquiries about data that support the findings of this study can be directed to the corresponding author.

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