

Dermoscopy of Primary Localized Cutaneous Nodular Amyloidosis

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Primary localized cutaneous nodular amyloidosis (PLCNA) is a rare skin condition typified by extracellular dermal deposition of amyloid proteins derived from immunoglobulin light chain L protein released by a localized infiltrate of plasma cells. It usually presents as solitary or multiple asymptomatic nodular lesions and its progression to systematic

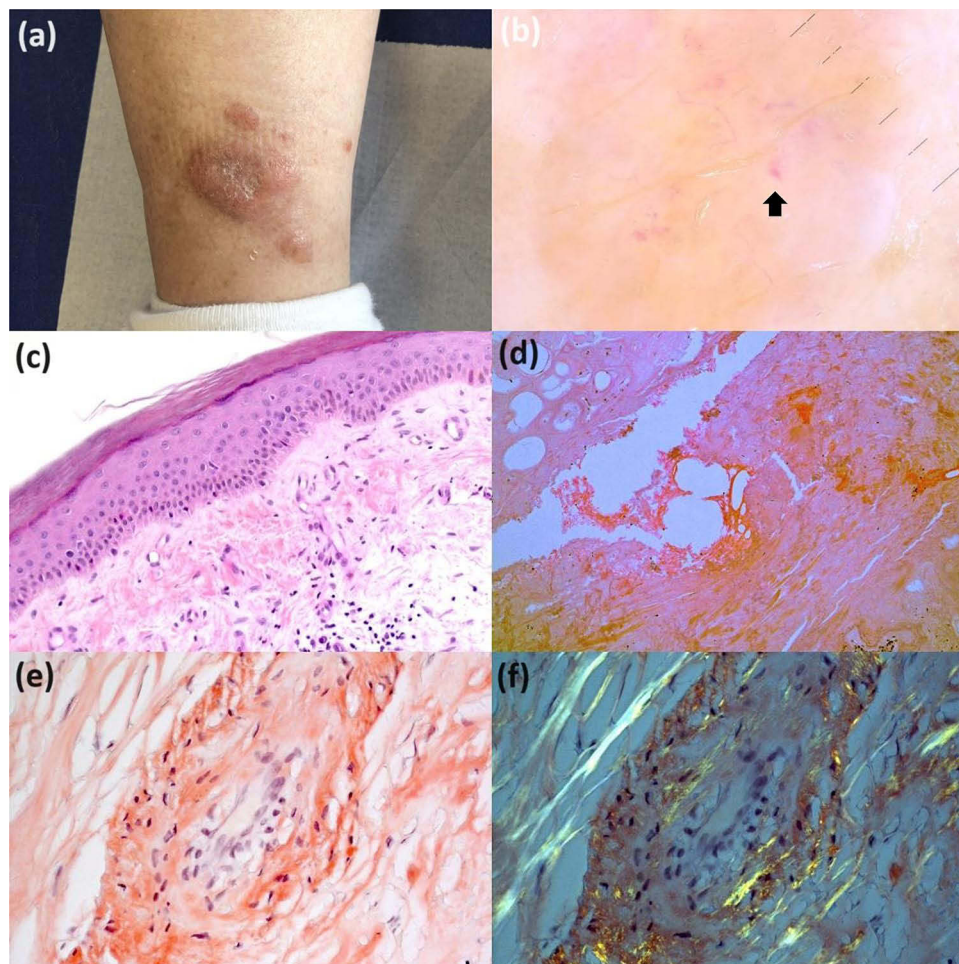


Figure 1 Pink-brown nodules and papules on the left leg (a). Polarized-light dermoscopy shows a diffuse Orange structureless, along with linear-curved vessels and purpuric (hemorrhagic) globules (arrow) (b). Histology shows deposition of pale eosinophilic amorphous substance in the superficial dermis (c) and hemorrhagic infiltrates at the dermo-hypodermal border (d); H&E x100. Positive staining for Congo red (e) and green apple birefringence under polarized light (f); x200.

Table 1 Previous Dermoscopy Reports on Dermoscopy of Primary Localized Cutaneous Nodular Amyloidosis

	Case	Site of Involvement	Comorbidities	Dermoscopy Description	International Dermoscopy Society Terminology	Histological Background
Atzori et al ¹	1	Left Leg	Limited cutaneous systemic sclerosis	- Structureless yellow background - Whitish scar strikes	- Yellow structureless areas (diffuse) - White lines	Not available
	2	Left Leg	Limited cutaneous systemic sclerosis	- Roundish waxy yellow blotches - Hemorrhagic background interspersed with fine telangiectasias - Hemorrhagic spots	- Yellow structureless areas (focal) - Purple globules - Vessels (not specified morphology)	- Nodular deposits of amorphous eosinophilic material in the dermis and subcutis - Peripheraeal patchy focal infiltrate of lymphocytes and plasma cells
	3	Both Legs	Limited cutaneous systemic sclerosis	- Structureless yellow background with whitish spots - Hemorrhagic halo - Elongated serpentine vessels	- Yellow structureless areas (diffuse) - White dots - Purple structureless areas (peripheral) - Linear-curved vessels	Not available
Rongioletti et al ²	1	Left leg	Not described	- Central orange-yellowish homogeneous area with elongated - Serpentine telangiectasias.	- Orange structureless areas (diffuse) - Linear-curved vessels	Acellular amorphous eosinophilic material on the dermis with peripheral plasma cells
Cheng et al ³	1	Left hallux nail bed	Diabetes mellitus	- Pink-orange background - Shiny white lines	- Orange and purple structureless areas (diffuse) - White lines	Eosinophilic amorphous extracellular deposits in the dermis, subcutaneous tissue, and blood vessel walls
Ferreira et al ⁴	1	Forth left toe	None	- OrangePink background - White shiny streaks	- Orange-pink structureless areas (diffuse) - White lines	Nodular deposits of hyaline and eosinophilic material, with spindle-shaped cells, in subcutaneous tissue and small vessels.
Sonagara et al ⁵	1	Left cheek	None	- Yellow-to-orange background with red linear and - Curved vessels - White dots and structureless white areas	- Orange structureless areas (diffuse) - Linear-curved vessels - White dots and structureless areas (focal)	Patchy nodular deposits of uniformly pink-stained amorphous material within the deep dermis and subcutaneous tissue

disease is estimated to occur in approximately 7% of the cases,¹⁻⁶ thus its recognition is of key importance to prompt diagnose any possible extra-cutaneous involvement. In this regard, dermoscopy has been shown to highlight some findings that may support PLCNA recognition, thereby facilitating the differential diagnosis with its clinical mimickers, mainly including cutaneous lymphomas and granulomatous dermatoses.¹⁻⁶

Here, we present a case of PLCNA with dermoscopic findings, also reviewing existing literature data on this topic and providing a homogeneous terminology according to the standardized dermoscopic criteria for non-neoplastic dermatoses released by the *International Dermoscopy Society*.⁷

A 75-year-old female presented with a 1-year history of asymptomatic pink-brown nodules on the posterior part of her left leg (Figure 1a). Medical history included systemic sclerosis and Hashimoto's thyroiditis. Dermoscopic assessment revealed a diffuse structureless orange area along with linear-curved vessels and few purple (hemorrhagic) globules/focal structureless areas (Figure 1b). Histological examination (Figures 1c and d) showed deposits of pale eosinophilic amorphous substance in the superficial dermis with hemorrhagic infiltrates at the dermo-hypodermal border; Congo red staining was positive (Figure 1e) and a green apple birefringence was evident under polarized light (Figure 1f). Therefore, a diagnosis of PLCNA was made.

According to the foregoing, the most represented dermoscopic feature in analyzed lesions of the present instance turned out to be a diffuse orange structureless area. This is in line with what described in previous seven cases reported in the literature (summarized in Table 1), that constantly showed orange/yellow structureless areas. Such a finding is likely related to the deposits of amyloid protein in the dermis and subcutis (the so-called "mass effect").⁸ Additionally, other features reported in prior instances included purple (hemorrhagic) background/dots (42.9% of cases), linear-curved vessels (42.9% of cases), and white dots/lines (71.4% of cases). Of note, in our patients we did not find white structures, probably as a result of the lack of dermal fibrosis, whereas we observed linear-curved vessels and purple (hemorrhagic) areas. Importantly, the detection of purple areas may be a significant clue in favor of PLCNA diagnosis compared to clinical mimickers showing orange color on dermoscopy, such as granulomatous dermatoses and skin lymphomas, that typically do not feature such a finding.^{8,9} Indeed, the presence of purple (hemorrhagic) structures is likely to be due to the peculiar vascular damage seen in skin amyloidosis with consequent erythrocytes extravasation. Obviously, comparative analyses are needed to confirm such observations.

Data Sharing Statement

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Compliance with Ethics Guidelines

This article is based on previously conducted studies and does not contain any new studies with human participants or animals performed by the authors. The patient in this manuscript has given informed consent to the publication of case details and institutional approval was not required.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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The authors report no conflicts of interest in this work.

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