CASE 27

Patient: A 57-year-old Thai female from Ayuthaya

Chief Complaint: Pruritic nodules on both legs for 3 years

Present Illness: The patient presented with a 3-year history of slightly pruritic, slow growing nodule on her shin that recently began to enlarge rapidly and developed numerous satellite nodules.

Past History: No underlying disease

Dermatological Examination (Figure 27.1-2)**:** Multiple discrete brownish hyperkeratotic papules coalescing to form plaques with discrete firm waxy yellow to brown pink nodules on both shins. Numerous tiny brownish macules distributed in a rippled pattern on the upper back.

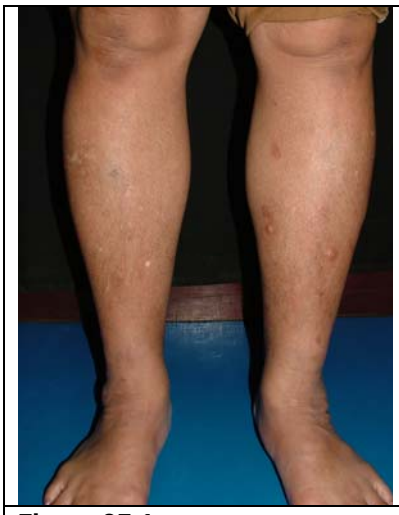

Figure 27.1

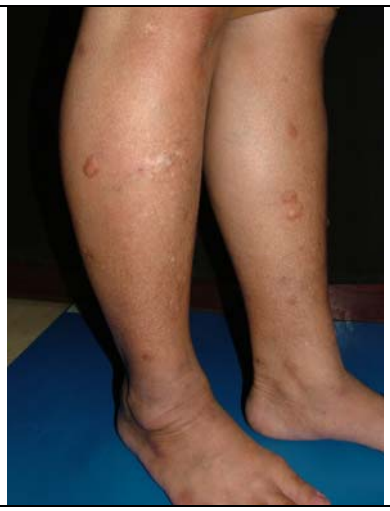

Figure 27.2

Physical Examination: unremarkable

Histopathology (S09-10991) (Figure 27.3)**:**

- Diffuse deposit of pale amphophilic material throughout the dermis with prominent around blood vessels, sweat glands and fat cells
- Patchy infiltrate of plasma within the dermis

Immunohistochemistry (Figure 27.4):

- Congo red staining: positive

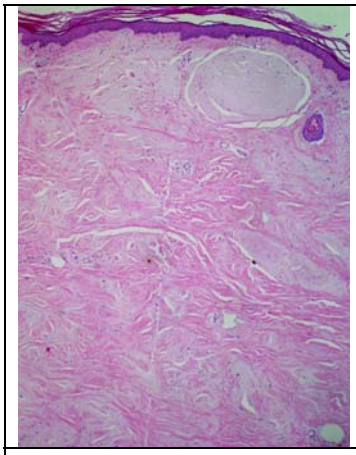

Figure 27.3, H&E 40x

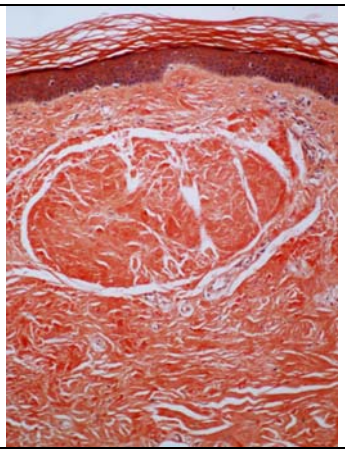

Figure 27.4, Congo red 100x

Investigations:

CBC:	Hct 37.1%, Plt 245,000
	WBC 5,320 /mm^3, N 61%, L 30%, Mo 6%
Chemistry:	BUN/ Cr: 23/0.6 mg/dl
LFT:	TP 74.7 g/L, Alb 36.5 g/L, ALP 83 U/L,
	AST 19 U/L, ALT 36 U/L, TB 0.3mg/dL, DB 0.1mg/dL
UA:	normal
Chest X-ray:	normal finding
Serum and urine electrophoresis:	normal
Abdominal fat pad aspiration:	pending

Diagnosis: Primary localized cuteneous nodular amyloidosis with macular and lichen amyloidosis

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Discussion:

Primary localized cutaneous amyloidosis has been classified to 3 types: macular, lichen, and nodular (tumefactive) according to the deposition of characteristic forms of amyloid fibril protein.[1]

Amyloid fibril protein	**Types of amyloidosis**	**Precursor substance**
Amyloid L	Primary localized cutaneous nodular amyloidosis	Immunoglobulin light chains(AL types) produced by local plasma cells
Keratin derivative	Macular and lichen amyloidosis	? Keratin filaments derived from degenerative keratinocytes

Primary localized cutaneous nodular amyloidosis (PLCNA) is a rare form of primary cutaneous amyloidosis that affects both sexs during middle age. It presents as waxy yellow-red nodules that are located preferentially on the lower extremities, face, scalp, and genitalia.[2] Some cases are associated with scleroderma and Sjogren's syndrome.[3] It has been reported that nodular amyloidosis may later develop paraproteinemia and overt systemic amyloidosis. However, there is an infrequent, 7%, progression of nodular localized lesions to systemic amyloidosis,[1,4] much lower than the 50% rate previously reported in the literature.[5] Patients with PLCNA should be investigated and followed-up to monitor for progression to systemic amyloidosis. If the results are normal, a good prognosis is advisable.

No previous literature reported coexisting of nodular with macular/lichen amylodosis is the same patient. The origin of two type are much different as epidermal damage is though to play a role in the development of lichen amyloidosis, whereas dermal amyloid L deposition which derive from immunoglobulin light chains produced locally by clonally expanded plasma cells is believed to be the source of nodular amyloidosis. However, recent reports document cases of PLCNA lacking clonality and occurring in common sites of trauma might explain co-occurrence between two types of localized cutaneous amyloidosis.[6] More data are needed before conclusions can be made regarding a causative role of epidermal damage in PLCNA.

Histologic diagnosis of nodular cutaneous amyloidosis is based on the presence of large hyaline eosinophilic masses of amyloid filling the entire dermis and occasionally the subcutaneous fat. Blood vessel walls are also involved. The histopathology does not permit the discrimination between localized forms and those that are manifestations of systemic amyloidosis, with the exception that in nodular amyloidosis there is usually an increased amount of infiltrating plasma cells. The deposits demonstrate an apple-green birefringence when viewd under polarized light with congo red stain, crystal violet and thioflavin T fluorescence.[7]

The management of PLCNA is difficult, as there is no consistently effective treatment. Cosmetic improvement can be achieved by surgical or shave excision,[8,9] successive electrodessication and curettage, dermabrasion, and carbon-dioxide laser.[10] As recurrences are common in contrast to lichenoid and macular amyloidosis, the least invasive procedure yielding cosmetically acceptable results is the best option.

References

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