

Primary Localized Cutaneous Nodular Amyloidosis with Macular and Lichen Amyloidosis: A Case Report.

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ABSTRACT:

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Primary localized cutaneous nodular amyloidosis (PLCNA) is a rare form of primary cutaneous amyloidosis that affects both sexes during middle age. There is an infrequent progression of nodular localized lesions to systemic amyloidosis. Here, we report coexisting of nodular amyloidosis with macular and lichen amyloidosis in the same patient which no previous literature has mentioned before. We also review the literatures regarding to rate of progression to systemic amyloidosis and discuss possible causes of co-occurrence between three types of localized cutaneous amyloidosis.

Key words: Nodular amyloidosis, lichen amyloidosis, macular amyloidosis

บทคัดย่อ :

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หน่วยโรคผิวหนัง ภาควิชาอายุรศาสตร์ โรงพยาบาลรามารักษ์ ต.มหาวิทาลัยมหิดล

Primary localized cutaneous nodular amyloidosis (PLCNA) เป็น cutaneous amyloidosis ที่พบน้อยมาก โดยเพศหญิงพบมากกว่าเพศชาย 2 เท่า พบมากในช่วงอายุ 50-70 ปี มีรายงาน nodular amyloidosis เปลี่ยนแปลงกลายเป็น systemic amyloidosis ได้ประมาณ 7% ภายในระยะเวลา 1-23 ปี ผู้เขียนได้รายงานผู้ป่วย nodular amyloidosis ที่พบ macular และ lichen amyloidosis ร่วมด้วย โดยทั้งนี้ยังไม่เคยมีรายงานการพบร่วมกันมาก่อน รวมทั้งได้รวบรวมข้อมูลการศึกษาอัตราการเปลี่ยนแปลงไปเป็น systemic amyloidosis และเหตุผลที่พบ primary cutaneous amyloidosis ทั้งสามชนิดร่วมกันในผู้ป่วยรายนี้

คำสำคัญ: อมิลอยด์โคซิส

Introduction

Primary localized cutaneous nodular amyloidosis (PLCNA) is a rare form of primary cutaneous amyloidosis that affects both sexes during middle age. There is an infrequent progression of nodular localized lesions to systemic amyloidosis. We reported coexisting of nodular amyloidosis with macular and lichen amyloidosis in the same patient which no previous literature has mentioned before. We also review the literatures regarding to rate of progression to systemic amyloidosis and discuss possible causes of co-occurrence between three types of localized cutaneous amyloidosis.

Case report

A 57-year-old Thai woman from Bangkok presented with 3-year- history of slightly pruritic, slow growing nodules on her shins that recently began to enlarge rapidly and developed numerous satellite nodules. She had no underlying diseases and was not taking any medications. Dermatologic examinations revealed discrete firm waxy yellow to brown- pink indurated subcutaneous nodules, vary in sizes on both shins (Figure 1), discrete brownish hyperkeratotic tiny papules coalescing to form plaques arranged in ripple pattern at posterior lower legs (Figure 2) and ill-defined brownish patch at upper back (Figure 3). Others physical examinations are unremarkable. Excisional

biopsy and histologic examination from a nodule on her right leg was performed.



Figure 1



Figure 2



Figure 3

Biopsy specimen revealed amorphous eosinophilic material compatible with amyloid deposition in dermis, subcutaneous tissue and in the vascular wall (Figure 4, 5). There are focal loose aggregation of lymphoplasmacytic cells with prominent Russel's bodies (Figure 6). Congo red stain shows birefringence apple green by polarized light microscope, consistent with AL amyloidosis.

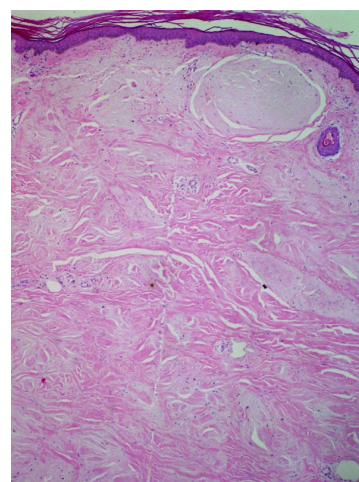


Figure 4

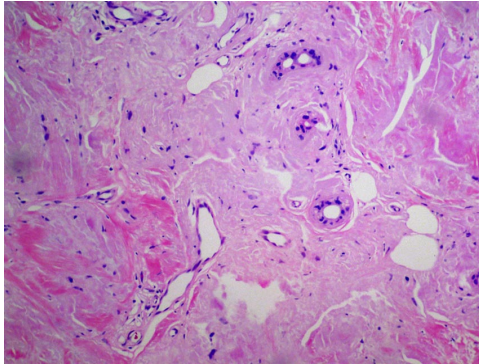


Figure 5

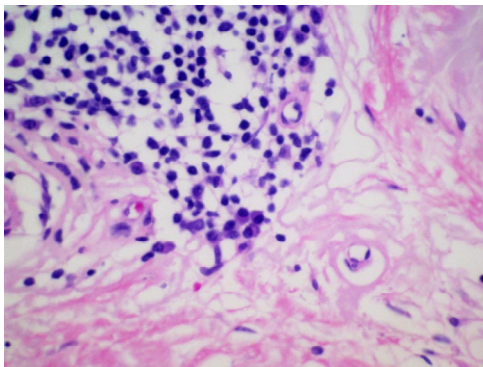


Figure 6

The laboratory investigations were performed to exclude the presence of an underlying condition which might contribute to systemic amyloidosis, including full blood count, erythrocyte sedimentation rate, plasma urea, creatinine, glucose and electrolytes, liver function tests, serum and urine electrophoresis, immunoglobulin and complement levels, auto-antibody profile, rheumatoid factor, urine microscopy, chest X-ray and abdominal fat pad biopsy.

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 (Table 1)

Table 1 Classification of primary localized cutaneous amyloidosis

Amyloid fibril protein	Types of amyloidosis	Precursor substance
Amyloid L	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 sexes 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, Sjogren's syndrome and rheumatoid arthritis.³ It has been reported that nodular amyloidosis may later develop paraproteinemia and overt systemic amyloidosis. Previous study reported that 40% of

patients presented with nodular amyloidosis also had monoclonal gammopathy which remained stable after follow-up. However, most recent case series demonstrate an infrequent, 7%, progression of nodular localized lesions to systemic amyloidosis^{1,4} (Table 2),

much lower than the 50% rate previously reported in the literature.⁵ Still, 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.

Table 2 Summary of follow-up cases of primary cutaneous nodular amyloidosis progressing to systemic amyloidosis

Reference	Total number of patients	Patients Progressed to systemic amyloidosis	Duration After diagnosis	*SPEP, UPEP	Laboratory Findings
Moon AO, et al 2003	16	1	1 year	Positive	Echocardiogram - normal
Woollons A, et al 2001	15	1	23 years	Positive	Baseline blood tests - normal
Vestey JP, et al 1994	1	1	7 years	Negative	CBC, ESR, LFT, chest X-ray, abdominal USG, EEG, Abdominal fat pad biopsy - normal
Brownstein MH, et al 1970	10	5	Not mentioned	Not mentioned	Not mentioned

*SPEP = Serum protein electrophoresis

UPEP = Urine protein electrophoresis

No previous literature reported coexisting of nodular with macular and lichen amyloidosis in the same patient. The pathogenesis of macular and lichen amyloidosis differs as epidermal damage is thought to play a role in its development 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. The presence of three types of cutaneous amyloidosis may be coincidental due to the high prevalence of macular and lichen amyloidosis in Asia. However, recent reports document cases of PLCNA

lacking clonality and occurring in common sites of trauma which 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 viewed under polarized light with congo red stain or dylon (cotton dye pagoda red No.9) stain. Amyloid can also be identified by sirius red, crystal violet, methyl 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 electrodesiccation and curettage, dermabrasion, and carbon-dioxide laser on solitary or lesions limited in amount.¹⁰ As recurrences are common in contrast to lichenoid and macular amyloidosis, the least invasive procedure yielding cosmetically acceptable results is the best option.

In our patient, findings from further investigations to exclude systemic involvement were negative, including results of serum and urine electrophoresis and abdominal fat pad biopsy. Indurated nodules were cosmetically disturbing to the patient hence they were removed by surgical excision.

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