Singapore Med J 2007; 48(6) : e165

Lichen amyloidosis in an unusual location

Jhingan A, Lee J S S, Kumarasinghe S P W

ABSTRACT

We report lichen amyloidosis occurring on the upper lip and nasolabial folds of a 61-year-old woman from Singapore. She had a past history of systemic lupus erythematosus, which was in remission for three years. There had been no lesions of lupus erythematosus in this area. Clinically, the lesions were skin-coloured, firm papules and our differential diagnoses included trichoepithelioma, papular sarcoid or lupus miliaris disseminatus faciei. Skin biopsy from one of the lesions showed amyloid deposits in the dermis which were Congo red stain positive. These deposits also showed apple green birefringence. Immunohistochemical staining of the amyloid deposits stained positive for cytokeratins (CK) 5 and 6, and negative for CK 14. The kappa and lambda stains were equivocal. Further investigations, including multiple myeloma screen and rectal biopsy, ruled out systemic amyloidosis. There was no other evidence of cutaneous amyloidosis on her limbs or trunk. She refused treatment for her lesions. This case highlights the commonly-seen form of primary localised cutaneous amyloidosis in an unusual location.

Keywords: amyloid proteins, amyloidosis, keratin, lichen amyloidosis, lip lesions

Singapore Med J 2007;48(6):e165–e167

INTRODUCTION

Amyloidosis is a group of rare conditions characterised by abnormal extracellular tissue deposition of one of a family of biochemically-unrelated proteins that share certain characteristic staining properties, including apple-green birefringence of Congo-red stained preparations viewed under polarised light and a fibrillar ultrastrucutre.[1] The amyloid is now classified based on the biochemical nature of amyloid fibril proteins rather than clinicopathological features.[2] We report a case of lichen amyloidosis (LA), which is the most commonly-seen form of primary localised cutaneous amyloidosis, occurring in an unusual location.

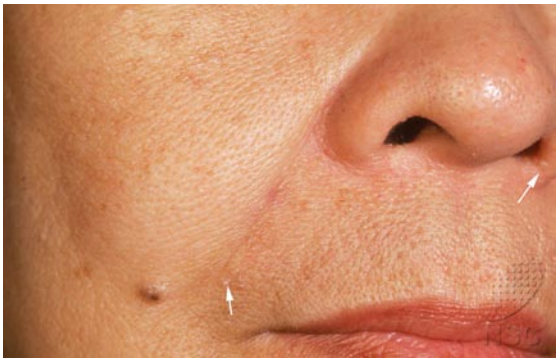

Fig. 1 Photograph shows discrete skin-coloured papules on the upper lip and left nostril (arrows).

CASE REPORT

A 61-year-old Chinese woman presented with a two-year history of asymptomatic, skin-coloured papules on the upper lip and nasolabial folds. In 1998, she had been diagnosed as having mild systemic lupus erythematosus (SLE) with leucopenia and arthritis of her interphalangeal joints and knees. Her white cell count was 3.1 cells/mm^3, antinuclear antibody (ANA) was positive at a titre of 1/640, speckled, lupus erythematosus cells, anti Ro and double-stranded DNA were all positive. She had no skin lesions at that time. She was started on oral prednisolone, which was gradually tapered and stopped in April 2002, and she has been in remission over the last three years. Physical examination revealed 15–20, 1–2 mm skin-coloured, discrete, firm papules on upper lip, right nasolabial fold and few new papules on left nasolabial fold (Fig. 1). The initial clinical impression was that of trichoepithelioma, papular sarcoid or lupus miliaris disseminatus faciei.

Histology showed expanded dermal papillae and elongated reteridges. They showed pink-stained globules and amorphous material (Fig. 2). A few compressed capillaries, and slight melanin, were present. Congo red stain was positive in these deposits (Fig. 3) and apple green birefringence was seen. A few apoptotic epidermal bodies were present in the basal layer. Solar elastosis was seen in the dermis. Appendages were normal. A histopathological diagnosis of dermal

National Skin Centre,
1 Mandalay Road,
Singapore 308205

Jhingan A, MBBS, DVD
Visiting Fellow

Lee JSS, MMed, MRCP, FAMS
Associate Consultant

Kumarasinghe SPW, MBBS, MD, FAMS
Senior Consultant

Correspondence to:
Dr. A Jhingan
Tel: (65) 6253 4455
Fax: (65) 6253 3225
Email: anjali@nsc.gov.sg

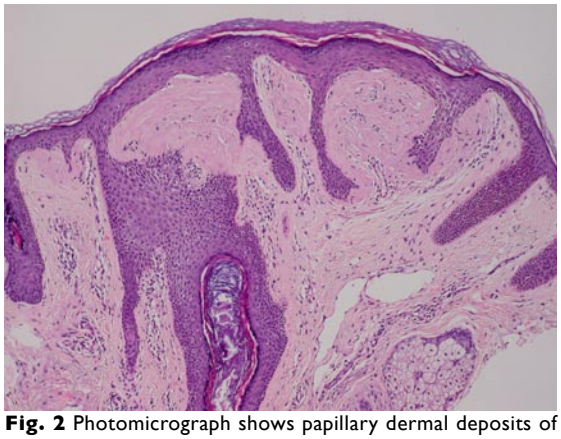

Fig. 2 Photomicrograph shows papillary dermal deposits of pink, amorphous amyloid (Haematoxylin & eosin, x 100).

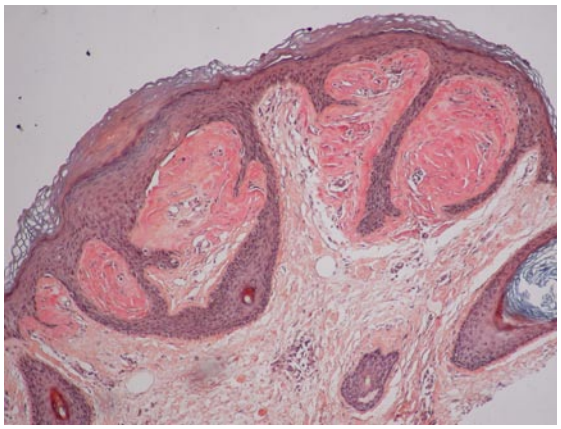

Fig. 3 Photomicrograph shows amorphous papillary dermal deposits stained red (Congo, x 100).

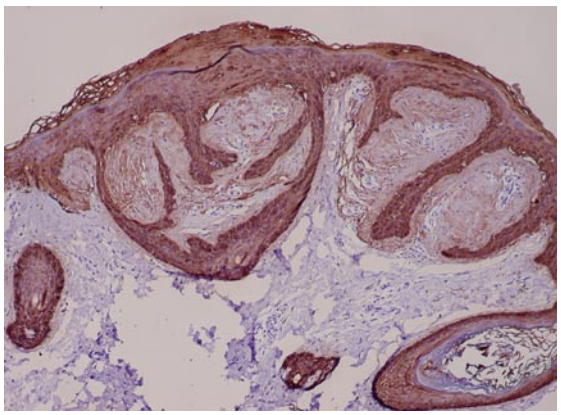

Fig. 4 Photomicrograph shows amorphous papillary dermal deposits stained positively for cytokeratins 5 and 6 (CK 5 and 6, x 100).

amyloidosis was made. Immunohistochemistry with the anticytokeratin antibodies showed positive staining of the amyloid deposits with cytokeratins (CK) 5 and 6 (Fig. 4) and negative with CK 14. Stainings for kappa and lambda light chains were equivocal.

Several investigations were carried out at this stage. The total white cell count was 3.90 cells/mm^3, red blood cell count was 3.76 cells/mm^3, haemoglobin level was 11.80 g/dL, and platelet count was 191 cells/mm^3. The differential count showed 41.1% neutrophils, 47.9% lymphocytes and 2% eosinophils. Erythrocyte sedimentation rate was 30 mm/h, urine microscopy was normal and ANA was negative. Investigations to exclude multiple myeloma, which included serum total protein, serum albumin, serum and urine protein electrophoresis were normal. No monoclonal band was detected on serum immunoelectrophoresis. Rectal biopsy showed no evidence of amyloid. At this stage, a diagnosis of LA was made. The patient refused treatment for her lesions.

DISCUSSION

Three types of primary cutaneous amyloidosis are recognised.[3] LA and macular amyloidosis are the most common and are associated with the deposition of amyloid in the papillary dermis. The third type, nodular, or tumefactive, primary localised cutaneous amyloidosis, in which the dermis, subcutis and blood vessel wall are diffusely infiltrated with amyloids, is very rare. It is believed to be derived from the local plasma cells. The exact characterisation of amyloid fibres in LA remains to be determined. Ultrastructural and immunohistochemical findings point towards degenerated keratin as the substrate.[4] LA is believed to be more common in persons of Chinese ancestry.[5] Patients reportedly range from 40 to 60 years of age. LA commonly presents as red-brown hyperkeratotic papules that coalesce into plaques, most commonly seen on the pretibial surfaces, arms, forearms, upper back and thighs. It is an intensely pruritic eruption though non-pruritic LA has also been described. Initial clinical differential diagnoses in our patient included trichoepithelioma, papular sarcoid or lupus miliaris disseminatus faciei. On detailed histological analysis, there was no evidence of trichoepithelioma, or any granuloma. Skin biopsy provided the definitive diagnosis. The amyloid in LA is present in the papillary dermis, usually within the dermal papillae.[6] LA can be distinguished from the macular form on histology, by the presence of hyperkeratosis, acanthosis, and larger deposits. The amyloid may be circumscribed, globular and could be in contact with basal cells at the basement membrane zone. Pigmentary incontinence, haemorrhage, and haemosiderin can also be seen in the papillary dermis. On electron microscopy, the amyloid of LA is composed of amyloid filaments, normal and degenerated tonofilaments, and lysosomes.[7] Direct

immunofluorescence in primary localised cutaneous amyloidosis reveals IgG, IgM, C3, and light chains, which are believed to be passively absorbed. Fibrils of lichen amyloid are believed to originate from epidermal keratinocytes. Immunohistochemical staining of the amyloid deposits reveal that monoclonal anticytokeratin antibodies are often reactive, suggesting its epidermal origin.[8] Various forms of treatment have been tried but none of them will eradicate the lesions. Options such as topical or intralesional corticosteroids, dimethyl sulfoxide,[9] retinoids,[10] ultraviolet therapy, calcipotriol, topical tacrolimus,[11] oral immunosuppressants, carbon dioxide laser vapourisation and dermabrasion have been used with varying success. For the relief of pruritus, sedating antihistamines have been found to be moderately beneficial. LA is typically benign and is limited to the skin. Cutaneous LA without associated systemic amyloidosis is common in Singapore.[12] This case highlights a commonly-seen form of primary localised cutaneous amyloidosis, in an unusual location, on the upper lip of a 61-year-old woman with a past history of SLE. There were no lesions of LA on her limbs and trunk and it was probably unrelated to SLE as there were no SLE lesions in this area in the past or at present. To our knowledge, this is the first reported case of LA on the upper lips.

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