Volume : 3 | Issue : 3 | March 2014 • ISSN No 2277 - 8160

Research Paper | Medical Science

Two Cases of Lichen Amyloidosis – A Rare Entity

Dr.Satish Arakeri Assistant Professor, Department of Pathology, DMWIMS, Naseera nagar, Meppadi(PO), kalpetta, Wayanad, Kerala, India.-673577

Dr.Priyadarshini D Assistant Professor, Department of Pathology, Shridevi institute of medical science and research hospital, Tumkur, Karnataka- 572106

ABSTRACT

Hyperpigmentation is the most common cosmetic skin complaint and distressing problem. Case report: First case: A 35 year male patient presented with macular lesions over extremities since 2 years. Second case: A 70 year male patient presented with multiple macular and papular rashes over back region since 3 years. Skin biopsy showed irregular acanthosis and hyperkeratosis. Papillary dermis showed eosinophilic amorphous material of amyloid . Congo red showed salmon pink color of amyloid. Polarising microscope showed apple green birefringence. Discussion: Lichen amyloidosis is a form of primary cutaneous amyloidosis in which amyloid is deposited in previously normal skin without any evidence of visceral involvement. Clinically it presents as pruritic small discrete hyperkeratotic hyperpigmented usually on the extensor surface of the lower limbs. It is treated by topical and intralesional corticosteroids, triamcinolone, dimethyl sulfoxide, keratolytic agents and dermabrasion

KEYWORDS : Lichen amyloidosis, pigmentation, congo red

Introduction:

Hyperpigmentation is the most common cosmetic skin complaint and distressing problem frequently encountered in females causing tremendous psychological impact due to their overanxious nature. It causes disfiguring lesions which can significantly affect a person's psychological and social well being, contributing to lower productivity, social functioning and self esteem.

Case report:

First case: A 35 year male patient presented with macular lesions over extremities since 2 years. The lesions were slowly increasing in size with increase in number also. These macules were brown to black pigmented in color.

Second case: A 70 year male patient presented with multiple macular and papular rashes over back region since 3 years. These lesions were black pigmented in color.

Skin biopsy of the cases was sent for histopathological examination. Epidermis showed irregular acanthosis and hyperkeratosis. Papillary dermis showed eosinophilic amorphous material of amyloid . Congo red staining had been done and showed salmon pink color of amyloid. Polarising microscope showed apple green birefringence.

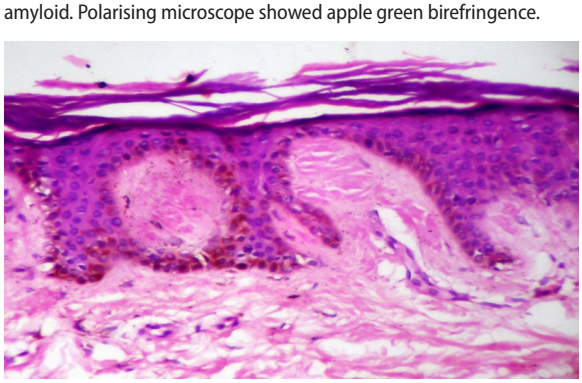

FIGURE 1: Skin biopsy irregular acanthosis, hyperkeratosis and with the papillary dermis showing eosinophilic amorphous material of amyloid (H&E stain 40X)

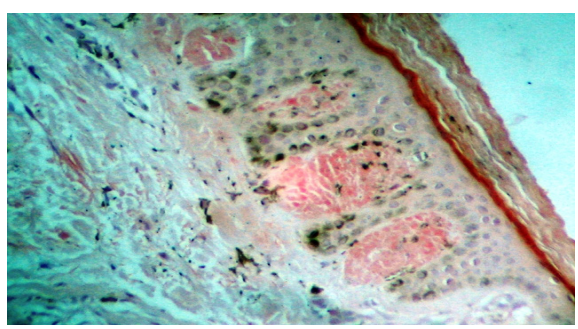

FIGURE 2: Congo red stain: amyloid in the papillary dermis shows a prominent "salmon pink colour"(40X)

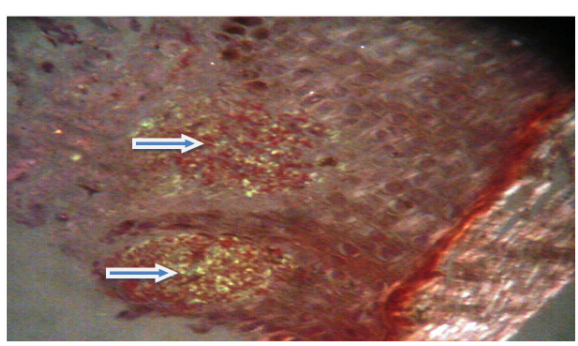

FIGURE 3: Congo red stain under polarised light showing "Apple green birefringence" in papillary dermis (40X)

Discussion:

The term "amyloid" was coined in 1854 by Virchow who was convinced by its resemblance to starch or cellulose. Rokitansky gave the first description of amyloidosis in 1842. Amyloid may accumulate as a result of a variety of different pathogenic mechanisms and the biochemical composition. Amyloid fibrils vary according to the clinico pathological type of amyloidosis. Amyloid fibrils are paired 7.5 to 10 nm, rigid, linear, non-branching, aggregated, hollow fibrils of indefinite length constitutes the bulk of amyloid deposits, regardless of clinicopathological type or the tissue involved, they are arranged in a loose meshwork. [1]

Lichen amyloidosis is a form of primary cutaneous amyloidosis in which amyloid is deposited in previously normal skin without any evidence of visceral involvement. It is most commonly seen in second and third decade of life with a slight female preponderance. Most of the pa-

tients are housewives. Majority of the patients have duration of illness of more than 1 year suggesting the chronicity of the disease. Friction due to scrubbing might be considered as a causative factor. Positive family history is noted in a considerable number of patients, indicating a possible role of genetic factors in the etiology of the disease.[2,3]

Clinically lesions consist of pruritic small discrete hyperkeratotic hyperpigmented, waxy papules in rippled pattern, usually on the extensor surface of the lower limbs. Pretibial area (shin) is the commonest site of involvement .Chronic irritation to skin has been proposed as an etiological factor. [2,3,6,7]

Cutaneous amyloidosis can be papular or lichenoid, macular and nodular or tumefactive types.

Lichen amyloidosis is the commonest type of localized cutaneous amyloidosis.

Pigmented macules with rippled pattern in macular amyloidosis and pigmented hyperkeratotic papules in lichen amyloidosis are the most common presentations and a combination of macular and lichen amyloidosis is seen in biphasic amyloidosis. Site most commonly involved in macular amyloidosis is extensor aspect of arm and anterior aspect of lower leg in lichen amyloidosis.[2,3,6,7]

Histopathology of such lesions show hyperkeratosis(100%), acanthosis(90%), papillomatosis (30%), hypergranulosis(16.7%) and elongation of rete ridges(5%). Amyloid deposits as uniform pink globules occupying dermal papilla in 95% cases. Amyloid deposits are visualized using Congo red in all 100% patients. Immunofluorescence shows the presence of IgM, C3 and IgA.[4,8]

Lichen amyloidosis has to be differentiated from various other clinical entities which closely resembles it. In lichen corneus hypertrophicus which probably represents a variant of lichen simplex chronicus, there are well-defined plaques composed of hemispherical hyperkeratotic, closely set, but usually discrete, intensely pruritic papules. The eruption has a predilection for ankles, shins, and calves. Clinically, the picture may be indistinguishable from that of lichen amyloidosis, differentiation is made on histological grounds.[2,3]

Lichen amyloidosis is often treated by topical and intralesional corticosteroids, triamcinolone, dimethyl sulfoxide, keratolytic agents and dermabrasion. In lichen amyloidosis a benefical effect has been noted after oral therapy with sedating antihistamines. In mild cases of lichen amyloidosis, potent topical corticosteroids in combination with a mild keratolytic agent such as salicylic acid may have added benefit. Intralesional injection of triamcinolone has been found beneficial . Topical use of dimethyl sulfoxide (DMSO) has been tried with varied success.[9]

Conclusion: Lichen amyloidosis is a rare entity should be suspected in any patient presented with chronic lichenoid lesions. Skin biopsy, Congo red stain with polarizing microscopy is essential to confirm the diagnosis.

REFERENCES

1. Kiboyashi H, Hashimoto K. Amyloidogenesis in organ- limited cutaneous amyloidosis: An antigenic identity between epidermal keratin and skin amyloid. The Journal of Investigative Dermatology 1983; 80: 66-72. | 2. Shenoi SD, Salim T, Balachandran C, Mehta RV. Lichen amyloidosis: A study of clinical, histopathologic and immunofluorescence findings in 30 cases. IJDVL 2005; 71(3): 166-169. | 3. Brownstein MH, Helwig EB. The cutaneous amyloidosis: Localized forms. Arch Dermatol 1970;102:8-19. | 4. Marchionini and John. Lichenoid and Poikiloderma like Amyloidosis. Arch Dermatol Syph. 1936;173:545-561. | 5. Al-Ratrout JT, Satti MB. Primary localized cutaneous amyloidosis: A clinicopathologic study from Saudi Arabia. Int J Dermatol. 1997;36:428-434. | 6. Wong CK. Lichen amyloidosis: A relatively common skin disorder in Taiwan. Arch Dermatol. 1974;110:438-440. | 7. Rajgopalan K and Tay CH. Familial lichen amyloidosis; report of 19 cases in 4 generation of a Chinese family in Malaysia. Br J Dermatol. 1972;87:123-129. | 8. Kibbi AB, Rubeiz NG. Primary localized cutaneous amyloidosis. Int J Dermatol. 1989;31(2):95-96. | 9. Das J, Gogoi RK. Treatment of primary cutaneous amyloidosis with cyclophosphamide. Indian Journal of Dermatol venereol Leprol 2003; 69(2):163-164. |