LICHEN AMYLOIDOSIS

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Amyloidoses are categorized into three major clinical subtypes: primary systemic, secondary systemic and organ-limited types. Both systemic and localized variants become more frequent with advancing age, and presentation before the age of 30 is rare. The amyloidoses are caused by deposition of abnormal proteins with at least 26 unrelated proteins are known to form amyloid fibril depositions.

Lichen amyloidosis accounts for approximately 10% of cutaneous amyloidoses. It is a cutaneous type of amyloidosis not associated with systemic amyloidosis but may occasionally be associated with other systemic conditions including: primary biliary cirrhosis, systemic lupus erythematosus and Sjögren’s Syndrome. Most often the cause of lichen amyloidosis is idiopathic but can be friction related.

Lichen amyloidosis is the most common type of cutaneous amyloidosis in Chinese individuals, usually affecting adults. Intensely pruritic hyperkeratotic papules that may coalesce to form gray to brown plaques with distribution on the anterior tibiae being a common site of involvement is a common presentation. However, spreading to extensor surfaces of the upper extremities and trunk may occur.

Laboratory testing for kidney and liver function as well as blood counts, creatinine clearance and measuring the level of protein in the urine should be part of the workup for all amyloidosis patients. For systemic variants of amyloidosis, further workup including bone marrow evaluation may be necessary.

Complications of lichen amyloidosis are usually related to pruritus with bleeding from excessive scratching. Rarely the lesions may be painful. The skin-limited lichen amyloidosis is not usually associated with any increase in mortality.

Chronic friction is a common etiological cause of lichen amyloidosis and treatment is usually directed at relieving the associated pruritus. Sedating antihistamines may help with relieving the itch, not because of their inherent antihistamine effects, but because of their inherent sedating qualities. Intralional steroids may be useful alone or in combination with other modalities including topical steroids, topical tacrolimus and even treatment with UVB phototherapy may help alleviate the pruritus. Systemic acitretin can be effective, but only as long as the patient continues the therapy. Surgical excision and dermabrasion of isolated lesions can be performed.