Read Item - Amyloidosis

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Abstract: Doctor's resource on Amyloidosis

Amyloidosis

Amyloid is a proteinaceous material that is deposited in tissues in a wide variety of conditions. The §-structure of the amyloid fibrils is resistant to proteolysis and so once deposited in tissues the amyloid persists indefinitely. The different proteins that make up the fibrils in different disorders account for the various clinical manifestations.

Systemic amyloidosis occurs when a serum acute phase protein is synthesised in excess in response to continued inflammation; or an immunoglobulin light chain component is produced in excess as a result of paraproteinaemia. The protein precipitates and is deposited perivascularly in a number of tissues and produces disease.

Cutaneous amyloid develops when an as yet unidentified protein derived from filamentous degeneration of the epidermis is deposited in the papillary dermis, however there is no involvement of internal organs.

In myeloma (or paraproteinaemia) related amyloidosis, the most characteristic lesions are yellow, waxy papules, which may be haemorrhagic. They occur on the face, particularly around the eyelids, and on the scalp. Nodules and plaques may also occur in systemic amyloidosis, but these are rare on the scalp. Diffuse infiltrates may mimic scleroderma.

Alopecia may be a conspicuous feature of systemic amyloidosis. Both a diffuse loss and a patchy loss of scalp hair can occur. Body hair may also be lost. Diffuse hair loss is associated with a loss of hair pigment and caused by cycling hairs failing to re-enter anagen (a prolonged telogen effluvium). Alopecia universalis has been described as a manifestation of occult systemic amyloidosis. Patchy loss is due to destruction of the pilosebaceous units by peri-appendageal deposition of amyloid. The proteinaceous material surrounds and compresses the hair follicle inducing atrophy and early loss of the hair.

Histology is diagnostic. In systemic amyloid there is deposition of amorphous masses of material perivascularly, periappendagealy and in the subcutaneous fat (amyloid rings). In cutaneous amyloid, deposits are found either in the subepidermal region (macular and lichen amyloidosis), or diffusely throughout the entire dermis and subcutis (nodular amyloid). The amyloid protein stains red with Congo red, and there is green birefringence when viewed with a polarised light.

Potent topical steroids are used for the lesions of cutaneous amyloid, but there is no treatment
known to be effective for systemic amyloid other than treatment of the underlying disorder.