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Head and neck amyloidosis: A clinicopathologic study of 15 cases

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Summary Amyloidosis refers to the idiopathic, extracellular deposition of fibrillar proteins, termed amyloid, in tissues. Although amyloidosis is a rare disease, the head and neck region has been reported as a frequent site of amyloid deposits, accounting for approximately 19% of reported amyloid cases in one review. Fifteen cases of head and neck amyloid, excluding the brain, with clinical follow-up were identified in the Surgical Pathology files from 1985 to 2005 at Emory University Hospital. The histopathology, histochemistry, and patient follow-up were reviewed. Nine men and six women with an age range of 18–76 years (mean 55.7 years) were identified. The initial clinical presentation was dependent on the site of amyloid deposits. The clinical types of amyloidosis included localized amyloid deposits in the larynx and tongue, plasma cell dyscrasia associated AL amyloidosis, and hemodialysis-associated amyloidosis. Secondary amyloidosis developed in one patient with carcinoid tumor.

Introduction

Amyloidosis refers to the idiopathic, extracellular deposition of fibrillar proteins (amyloid) in tissues, which, if extensive, can interfere with normal function and ultimately lead to death if vital organs are affected.[1] Diseases associated with this protein vary in severity and may be localized or systemic, inherited or acquired, innocuous or life threatening. More than 25 distinct biochemical forms of amyloid have been identified (Table 1).[2] Amyloid deposits occur in any tissue, either localized or diffusely.

Electron microscopy shows that all amyloid subtypes are composed of 7.5–10 nm wide linear, nonbranching tubular protein fibrils loosely arranged in a meshwork. However, extensive study of this

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Table 1 Classification of amyloidosis

Classification	Precursor protein	Associated diseases
Systemic amyloidosis		
AL	Immunoglobulin light chains, mainly λ type	Multiple myeloma, plasma cell dyscrasia, primary AL amyloidosis, may be localized
AA	Serum amyloid A	Chronic inflammatory conditions (RA, Crohns disease, malaria, TB); malignancy (Hodgkin lymphoma, GI, GU, and lung carcinoma); secondary AA amyloidosis
AH	Immunoglobulin heavy chain	Primary AA amyloidosis, myeloma associated, may be localized
Aβ_2M	β_2-microglobulin	Hemodialysis-associated amyloidosis
ATTR	Transthyretin	Familial amyloidotic neduropathies (mutant form), systemic senile amyloidosis (native form)
AApoI	Apolipoprotein AI	Familial systemic amyloidosis
AApoAII	Apolipoprotein AII	Hereditary renal amyloidosis
AApoAIV	Apolipoprotein AIV	Sporadic, associated with aging
AFib	Fibrinogen α-chain	Hereditary renal amyloidosis
ALys	Lysozyme	Familial visceral amyloidosis
Localized amyloidosis—central nervous system		
Aβ	AβPP	Sporadic Alzheimer disease, aging, Down syndrome
AprP	Prion protein	Sporadic Creutzfeldt-Jacob disease
ACys	Cystatin C	Hereditary cerebral hemorrhage with Amyloidosis (Icelandic)
ABri	ABriPP	Familial dementia, British
ADan	ADanPP	Familial dementia, Danish
Localized amyloidosis—ocular		
AGel	Gelsolin	Familial (Finnish) amyloid polyneuropathy, lattice corneal dystrophy
ALac	Lactoferrin	Familial corneal amyloidosis
AKer	Kerato-epithelin	Familial corneal dystrophies
Localized amyloidosis—localized		
AIPP	Islet amyloid polypeptide	Insulinoma, Type 2 diabetes
APro	Prolactin	Pituitary amyloid
AANF	Atrionatriuretic peptide	Atrial amyloidosis
AIns	Insulin	Iatrogenic
ACal	Calcitonin	Medullary thyroid carcinoma
AMed	Lactaherin	Aortic amyloidosis of elderly
A(tbn)[a]	tbn	Pindborg tumor

[a] A(tbn): to be named; waiting for a protein name.