

Primary breast amyloidosis case report and literature review

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Abstract

Background: Primary breast amyloidosis is a rare benign disease characterized by the accumulation of insoluble amyloid protein.

Case report: A 50-year-old female with a past medical history of Sjögren's syndrome has newly diagnosed left breast amyloidosis. She has had left breast abnormalities since 2015 with follow-up mammogram showing slightly changed left breast abnormality. Subsequent biopsy showed Congo red positive amyloid deposition. Mass spectroscopy LC MS/MS detected a peptide profile consistent with immunoglobulin-associated amyloid deposition. She also has a one-year history of bilateral thigh rash with skin biopsy confirmed cutaneous T cell lymphoma. No systemic amyloidosis was detected.

Literature review: By searching Pubmed using "breast and amyloidosis, 98 cases were found in the last 10 years with 1 male and 97 females (38 - 85 years). Additional breast findings include Non-Hodgkin's lymphoma (B/T cell lymphoma, mucosa-associated lymphoid tissue lymphoma, extranodal marginal zone lymphoma, lymphoplasmacytic lymphoma, n=49); plasma cell proliferative disorder/plasmacytoma (n=11); breast cancer (invasive carcinoma or ductal carcinoma in situ, n=4); and chronic lymphoid leukemia (n=1). The most common amyloid type is AL Kappa (n=18); followed by AL Lambda (n=14); AH/AL (n=2); Amyloid transthyretin (n=2) and Iatrogenic insulin-derived amyloidosis (n=2). Associated autoimmune diseases include primary Sjögren's syndrome (n=7), CREST syndrome (n=1), and type 1 diabetes mellitus (n=1). No patient needed systemic chemotherapy.

Summary and conclusion: Different than systemic amyloidosis, breast amyloidosis is a benign disease. Since it can coexist with malignancy disease, appropriate workup must be thorough but not overly extensive.

Parameters		No. of patients
Gender /age	Female (38-85yo)	97[1]
	Male (not clear)	1[1]
Additional breast findings	Non-Hodgkin's lymphoma, including B/T cell lymphoma, MALT lymphoma /Extranodal marginal zone lymphoma/Lymphoplasmacytic lymphoma	49[1-3]
	Plasma cell proliferative disorder/plasmacytoma	11[1]
	Intraductal carcinoma, ductal carcinoma in situ, invasive ductal, papillary and lobular carcinoma	4[2]
	Chronic lymphoid leukemia	1[1]
Type of Amyloid	AL Kappa	18[1, 4, 5]
	AL Lambda	14[4, 6]
	AH/AL	2[1, 7]
	Amyloid transthyretin	2[4]
	Iatrogenic insulin-derived amyloidosis	2[4, 8]
Associated disease	Unspecified Autoimmune disease/ Sjogren's syndrome	7[9-11]
	Remote dominant plasma cell myeloma	1[12]
	Monoclonal gammopathy of undetermined significance	1[6]
	CREST syndrome	1[13]

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