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Supplement to IgG4-related Breast Disease: Review of the Literature

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Table 1. Clinical, Imaging, and Pathological Data of IgG4-BD Cases Reported in the Literature.

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 1 (24/F) ¹⁴	Pain and redness in right breast	US: Breast abscess	Vacuum and excisional	142	Focal architectural distortion, dense inflammatory infiltrate with dense sclerotic stroma. No granulomas	45	Prednisone 1 mg/kg MTX 15 mg/week	Resolution within 3 months
Pt. 2 (70/F) ¹⁵	Axillary lymph nodes, soft egg-sized tender mass in left breast	MG: Breast lump US: Hypochoic mass with irregular border and poor Doppler signal MRI: Breast mass—suspected tumor	Yes, type not defined	—	Mixed T and B lymphocytic and plasma cell infiltration with interstitial fibrosis	40	Prednisone 10 mg/day	Lesion resolution stable during 4-year follow-up
Pt. 3 (56/F) ¹⁶	Palpable non-painful lump in left breast	US: Ill-defined hypochoic mass with low vascularity; no shadowing or calcification MRI: Non-mass enhancement	Yes, type not defined	—	IgG4-related sclerosing mastitis	—	—	—

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 4 (52/F) ¹⁷	Asymptomatic left breast lump on screening mammography	MG: Small circumscribed opacity US: Small solid avascular hypoechoic nodule with hyperechogenic tiny foci	Core	131	Scattered stromal cells with chronic dense inflammatory infiltrates and prominent IgG4 expression	—	Excision	—
Pt. 5 (71/F) ¹⁸	Asymptomatic left breast mass on CT (screening for IgG4-RD because of eye involvement)	MG and US: Breast mass—suspected tumor MRI: Bordered breast mass with early enhancement—suspected tumor	Excision of large nodules	1430	Dense lymphoplasmacytic infiltration with CD20 ⁺ B cells and CD3 ⁺ T cells	76	Prednisone 22.5 mg/day, tapered to 5 mg/day as maintenance	Improved within 3 months; stabilized after 1 year
Pt. 6 (84/F) ¹⁹	Palpable unilateral non-painful lump (unknown side)	MG: Breast mass without calcifications or architectural distortion US: Well circumscribed round hypoechoic mass with vascular flow on Doppler	Excision of large nodules	—	Fibro-inflammatory lesion with dense lymphoplasmacytic infiltration, numerous plasma cells, eosinophils	50	Mass excision	—
Pt. 7 (56/F) ²⁰	Palpable bilateral non-painful lump	MG: Mass-like densities, both breasts US: Irregular ill-defined hypoechoic masses PET: Mildly increased metabolic activity	Core biopsy	Elevated	Extensive stromal fibrosis with robust IgG4 ⁺ plasma cells in breast and dermal tissue	40	Steroids	Orbital disease resolution; stable breast mass
Pt. 8 (56/F) ²¹	Recurrent mastitis (unknown side)	—	Excision of large nodules	920	IgG4 infiltrates	—	Prednisone 40 mg/day tapered to 10 mg/day maintenance therapy	Symptom resolution; stable after 1 year
Pt. 9 (41/F) ²²	Asymptomatic right breast lump on screening mammography	MG and US: Small mass	Biopsy	39.4	Sclerotic stroma with dense lymphoplasmacytic infiltrates	43	Observation	—
Pt. 10 (54/F) ²³	Palpable non-painful left breast lump	US: Irregular ill-defined hypoechoic masses CT: Axillary nodes enlargement	Biopsy	3320	Mixed B, T, and plasma cell infiltrates with interstitial fibrosis	40	Prednisone 30 mg/day for 4 weeks and maintenance therapy	Decrease in breast mass

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 11 (48/F) ²⁴	Palpable bilateral non-painful lump	MG: Breast nodules US: Hypoechoic nodules with lobular margins MRI: Multiple lesions, both breasts	US-guided biopsy	650	Dense mass, lymphoplasmacytic infiltrates, prominent stromal sclerosis, loss of breast lobules; mostly B-CD20 ⁺ and T CD3 ⁺ lymphocytes	65	Excision of large nodules	Stable state
Pt. 12 (51/F) ²⁴	Palpable right breast non-painful lump	MG: Breast nodules	Core biopsy	3900	Dense mass, lymphoplasmacytic infiltrates, prominent stromal sclerosis, loss of breast lobules; mostly B-CD20 ⁺ and T CD3 ⁺ lymphocytes	85	Excision of large nodules	Stable state
Pt. 13 (37/F) ²⁴	Palpable right breast non-painful lump	MG: Breast nodules	Core biopsy	—	Dense mass, lymphoplasmacytic infiltrates, prominent stromal sclerosis, loss of breast lobules; mostly B-CD20 ⁺ and T CD3 ⁺ lymphocytes. Phlebitis	82	Core needle biopsy	Breast mass resolved, lymphadenopathy unchanged
Pt. 14 (54/F) ²⁴	Palpable right breast non-painful lump	MG and US: Fibro-adenoma-like lesions	Performed	—	Dense mass, lymphoplasmacytic infiltrates, prominent stromal sclerosis, loss of breast lobules; mostly B-CD20 ⁺ and T CD3 ⁺ lymphocytes	49	Excision of large nodules	Stable state
Pt. 15 (62/F) ²⁵	Painful right breast mass	US: Hypoechoic mass	US-guided	—	Granulomatous lobular mastitis with giant cells, sclerosis, and IgG4 ⁺	—	—	—
Pt. 16 (66/F) ²⁵	Painful right breast mass	US: Ill-defined hypoechoic mass	Surgical excision	189	—	—	—	—
Pt. 17 (49/F) ²⁶	Multiple painless masses in left breast	US: Hypoechoic uncompressible mass, surrounding architecture disturbance without acoustic shadowing	Biopsy excision	390	Sclerotic lesions with low cellularity and plasma cell-rich inflammatory infiltrate	51	Steroids	Partial regression
Pt. 18 (47/F) ²⁷	Asymptomatic breast lump (unknown side)	—	—	140	Reactive lymphoid follicles, fibrosis, elevated eosinophils and plasma cells, no phlebitis	30	—	Recurrent orbital mass
Pt. 19 (61/F) ²⁸	Asymptomatic right breast lesion on planned PET-FDG	MG and US: Performed, no description PET-CT: Increased metabolic activity in the breast (planned exam for lymphoma)	—	321	Sclerotic lesions with low cellularity and plasma cell-rich inflammatory infiltrate: lymphocytes, plasma cells, eosinophils, histiocytes. Carcinoma <i>in situ</i>	85	Excision	—

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 20 (59/M) ²⁸	Asymptomatic left breast lesion on planned total body CT	MG: Multinodular irregular mass CT: For prostatic cancer assessment, breast mass	–	–	Stromal sclerosis with dense plasma cells (>30 HPF) infiltrate, positive for IgG4	–	Excision	–
Pt. 21 (48/M) ²⁹	Asymptomatic right breast lump	MG: Breast mass involving pectoral muscle with skin thickening US: Heterogeneous mass with mild vascularity	–	1220	Fibrotic tissue with storiform pattern and dense lymphoplasmacytic infiltrates, several eosinophils, no granuloma	51	Excision	–
Pt. 22 (66/F) ³⁰	Asymptomatic left breast lump	MG: Breast mass—suspected tumor	–	179	Storiform fibrosis, dense lymphoplasmacytic infiltrates, obliterative phlebitis	63.9	Excision	–
Pt. 23 (45/F) ³⁰	Asymptomatic right breast lump	MG: Breast mass—suspected tumor	–	308	Storiform fibrosis, dense lymphoplasmacytic infiltrates, obliterative phlebitis	67.3	Excision	–
Pt. 24 (37/F) ³¹	Asymptomatic breast lump (unknown side)	MG: Breast lesion	Performed	166	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis, giant cells	75.1	–	–
Pt. 25 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	99.3	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	47.5	–	–
Pt. 26 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	68.3	Dense lymphoplasmacytic infiltrates, storiform fibrosis	45.1	–	–
Pt. 27 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	64	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	74.1	–	–
Pt. 28 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	52.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis, histiocytes	57	–	–
Pt. 29 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	48.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis	59.6	–	–
Pt. 30 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	42.3	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis, histiocytes	32	–	–
Pt. 31 (–/–) ³¹	Asymptomatic breast lump (unknown side)	–	–	41.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis	40.2	–	–

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 32 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	41	Dense lymphoplasmacytic infiltrates, storiform fibrosis	40.5	—	—
Pt. 33 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	31	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	62	—	—
Pt. 34 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	28.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis, histiocytes	18.9	—	—
Pt. 35 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	27.3	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	49.4	—	—
Pt. 36 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	26.3	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	61.2	—	—
Pt. 37 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	19.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	34.1	—	—
Pt. 38 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	17.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis, histiocytes	49.5	—	—
Pt. 39 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	16.7	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis	42	—	—
Pt. 40 (-/-) ³¹	Asymptomatic breast lump (unknown side)	—	—	12	Dense lymphoplasmacytic infiltrates, storiform fibrosis, obliterative phlebitis, histiocytes	46.2	—	—
Pt. 41 (82/F) ³²	Asymptomatic right breast lump	MG: Ill-defined focal asymmetric infiltrates, no calcifications US: Ill-defined heterogeneous mass	Core biopsy	—	Polyclonal T and B cell infiltrates, macrophages, plasma cells, histiocytes, lymphoplasmacytic infiltrates with fibrosis and sclerosis	—	—	—
Pt. 42 (65/F) ³³	Asymptomatic right breast lump	—	Excisional biopsy	150	Robust lymphocytes and plasma cells, diffuse fibrosis, IgG4 ⁺ cells 172/HPF, few obliterative phlebitis with concomitant features of Rosai- Dorfman disease	82	Excision	No recurrence

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 43 (78/F) ³³	Asymptomatic right breast lump	PET: Right breast mass and right axillary lymph nodes with high FDG uptake	Excision	18	Robust lymphocytes and plasma cells, diffuse fibrosis, IgG4 ⁺ cells 118/HPF, few obliterative phlebitis with concomitant features of Rosai- Dorfman disease	53	Excision	No recurrence
Pt. 44 (54/F) ³⁴	Asymptomatic right breast lump	MG: Irregular-shaped, dense lesion with indistinct margins US: Low-echoic lesions CT: For eyelids assess- ment, breast mass noted MRI: Segmental enhancement	Core needle biopsy	217	Irregular stromal cell proliferation associated with severe lymphoplasmacytic infiltration, adenosis, and intraductal papilloma surrounded by fibrosis; diffusely distributed IgG4 ⁺ plasma cells	—	Prednisone 40 mg/day tapered to 10 mg/day maintenance therapy	No recurrence
Pt. 45 (43/F) ³⁵	Asymptomatic right breast lump	US: Low-echoic breast mass with abundant vascular signal PET: Right breast mass and right axillary lymph nodes with high FDG uptake MRI: Irregular shaped con- trasted mass with enhancement	Extraction	517	Dense fibrosis with abundance of plasma cells	70	Prednisone 30 mg/day tapered to 5 mg/day maintenance therapy	No recurrence
Pt. 46 (62/F) ³⁶	Asymptomatic right breast lump	MG: Nodule, right lower portion US: Hyperechoic nodule in right lower portion	Performed for diag- nosis and surgical extraction	—	Large polygonal histiocytes with zonal proliferation pattern, lymphoplasmacytic cells with stromal fibrosis with concomitant features of Rosai-Dorfman disease; positive reactions for IgG and IgG4 in fibrotic stroma	56.7	Surgical extraction	No recurrence
Pt. 47 (69/F) ³⁷	Asymptomatic left breast lump	MG: Bilateral diffuse calcifications and multiple circumscribed masses in the medial left breast US: Multiple masses in the left breast and chest wall; abnormal left axillary lymph nodes MRI: Multiple heteroge- neous enhancing masses in left breast and chest wall surrounding and involving multiple ribs	US-guided	191	Mixed inflammatory infiltrate with background fibrosis; no staining for IgG4 was performed	—	—	—

Pt. # (age, y/sex) ^{ref.}	Presentation	Imaging	Biopsy	Serum IgG4 (mg/dL)	Histopathology	% IgG4 ⁺ /IgG ⁺	Treatment	Outcome
Pt. 48 (46/F) ³⁸	Indurative left breast lump	MG and US: Ill-defined nodule MRI: Breast mass with enhancement	Excisional	185	Irregular proliferation of stromal cells associated with severe lymphoplasmacytic infiltration, obliterative phlebitis, and eosinophils; many plasma cells within the lesion immunohistochem- ically positive for IgG4	—	—	—

—, not known; CT, computed tomography; F, female; FDG, fluorodeoxyglucose; HPF, high-power field; M, male; MG, mammography; MRI, magnetic resonance imaging; MTX, methotrexate; PC, plasma cells; PET, positron emission tomography; US, ultrasonography.