

TABLE 2.

Other skin disorders associated with monoclonal gammopathies

Disease	Monoclonal gammopathy	Clinical	Pathology	Other
Strong association				
Scleromyxedema	IgG λ	Widespread, symmetric eruption of 2–3 mm firm, waxy, papules often on dorsal hands, extensor extremities, face Diffuse skin thickening	Mucin in the upper and mid-reticular dermis Thickened dermis with increased collagen and increased (stellate) fibroblasts	Deep longitudinal glabellar furrows
Scleredema, group III with associated monoclonal gammopathy	IgG $\kappa > \lambda$	Firm non-pitting edema of the trunk, face, and neck	Thickened reticular dermis with mucin between collagen bundles; lacks increase fibroblast density	Other groups: Group I: after acute respiratory tract infection Group II: associated with diabetes
Necrobiotic xanthogranuloma	IgG $\kappa > \lambda$	Yellowish-red, indurated plaques with telangiectasia and atrophy +/- ulceration Often peri-orbital	Palisading granulomatous collections of histiocytes, Touton-type giant cells, and bizarre-appearing giant cells	May have multiple myeloma or cryoglobulinemia as well
Plane xanthoma, diffuse normolipemic (Group II, type 2)	IgG	Yellowish patches and plaques Involves eyelids, neck, upper trunk, buttocks, flexures	Foamy histiocytes in upper dermis, often clustered around vessels	Often have multiple myeloma
	Type III	Polyclonal IgG or IgM		
Schnitzler's syndrome	IgM κ	Chronic urticaria	Neutrophilic infiltrate within venules and perivascular +/- leukocytoclasia, fibrinoid necrosis	Bone pain, fever, hyperostosis
Glomeruloid Hemangioma	Variable	Polypoid, angiomatous papules Often eruptive and multiple	Dilated vascular spaces lined by flat endothelial cells, resembles renal glomerulus	POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy, and Skin changes)
Weak association				
Pyoderma Gangrenosum	IgA > other	Ulceration with violaceous, raised, undermined borders	Diffuse neutrophilic infiltrate, often with follicular involvement	
Sweet syndrome (Acute febrile neutrophilic dermatosis)	Variable	Tender, erythematous papules and plaques	Dermal neutrophilic infiltrate	