Appendix 1 (as supplied by the authors): Key features on skin biopsy in the main diffuse sclerosing skin conditions[1-3]

Condition	Key features on skin biopsy
Diffuse systemic sclerosis (dcSSc)	Excessive deposition of extracellular matrix proteins, leads to fibrosis of the reticular (deep) dermis and the subcutaneous fibrous trabeculae. In the early stages, collagen bundles are seen within the reticular dermis, which appears pale, homogenous, and edematous. Lymphocytic infiltrate is notable in a perivascular distribution, in between the collagen bundles, in the subcutaneous fat, encasing the sweat glands and also involving the dermal blood vessels. Subsequently endothelial proliferation leads to complete occlusion of the blood vessels. With disease progression, the involved skin becomes more avascular and inflammation decreases. The overlying epidermis becomes atrophic. Also, pilosebaceous units and eccrine glands disappear, collagen bundles become more closely packed, and the rete ridges get effaced.
Scleredema	Histopathologically the different subtypes of scleredema are indistinguishable. Epidermis is normal. There is noticeable thickening of the reticular dermis, due to increased thickened collagen bundles, which also extend into the subcutis. The collagen fibers are swollen and separated from one another, interspersed with deposits of mucin. There is accumulation of acid mucopolysaccharide (hyaluronic acid), predominantly in the deep dermis, with concurrent dermal sclerosis. Increased mucin deposition can be highlighted with special stains, e.g., colloidal iron and Alcian blue. Inflammatory cells are sparse. Elastic fibers are reduced and fragmented. Unlike scleroderma, dermal appendages are preserved. Also, unlike scleromyxedema, there is an absence of fibroblast proliferation.
Nephrogenic systemic fibrosis (NSF)	Early in the course there is diffuse dermal proliferation of epithelioid, spindled and dendritic fibrocytes, increased collagen fibers, and increased interstitial mucin in the dermis that may infiltrate the subcutaneous septa. Giant cells are occasionally present. The dermal fibrocytes, recruited from the circulating fibrocyte pool, co-express CD34 and procollagen-1. Interstitial mucin eventually disappears over time. Unlike scleromyxedema, NSF always involves the lower dermis often extending to the pannicular septae, whereas scleromyxedema tends to be restricted to the upper half of the dermis.
Diffuse fasciitis with eosinophilia (DFE)	Characteristic findings on a full thickness excisional biopsy include chronic perivascular lymphoplasmacytic inflammation along with histiocytes, mast cells and eosinophils infiltrating the deep dermis, fascia and superficial muscle, but sparing the epidermis, papillary dermis and the adnexa. Thickening of the deep fascia results from sclerosis and fibrosis. Subjacent muscle shows mild atrophy, and a chronic, predominantly perivascular inflammation.
Scleromyxedema	There is superficial to mid-dermal mucin deposition along with fibroblast proliferation and fibrosis. The pannicular septae are never involved. Pathology tends to be restricted to the upper half of the dermis only.

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