Lichen Sclerosus ET Atrophicus
Lichen sclerosus (LS) encompasses the disorders known as lichen sclerosus et atrophicus, balanitis xerotica obliterans (LS of the male glans and prepuce), and kraurosis vulvae (LS of the female labia majora, labia minora, perineum, and perianal region). Lichen sclerosus is an inflammatory dermatosis of the skin and mucous membranes. It may be asymptomatic or symptomatic, and often the only, site of involvement. Extragenital lesions may occur with or without coexisting genital lesions.

Lesions of LS are characterized by white polygonal papules that coalesce to form plaques. Comedo-like...
In male patients, involvement of the glans and prepuce often results in phimosis. Although the literature...

In female patients, contiguous involvement of the labial, perineal, and anal areas has been described cli...
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or "keyhole" lesions (165). Many cases of childhood LS in girls resolve by menarche (166). If lesions persist, they may cause pruritus, which is common in the vulvar region. The premalignant potential in LS has been debated extensively and remains ill defined. The most recent large studies have shown that the risk of malignancy in LS is low. However, the long-term follow-up of patients with lichen sclerosus et atrophicus of the vulva is advisable. Of interest, lesions of LS may koebnerize (be provoked by trauma) as well as coexist with morphea (167). In extensive cases of morphea, lichen sclerosus et atrophicus may become superimposed on some of the lesions. It is then best...
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recognized by finding pale superficial dermal collagen, as compared with hypocellular compacted deep collagen. Cases of both extragenital and genital LS have also been reported in the setting of graft-versus-host disease.
Histopathology.

The salient histologic findings in cutaneous lesions of lichen sclerosus et atrophicus include (a) hyperkeratosis with acanthosis, (b) spongiosis, (c) edema and homogenization of the collagen in the upper dermis, and (d) an inflammatory infiltrate in the mid-dermis.
The hyperkeratosis is so marked that the horny layer is often thicker than the atrophic stratum malpighii, which may be irregularly proliferated. In such proliferations, hydropic degeneration of the basal cells usually is pronounced.

Keratotic plugging of appendageal ostia is often associated with atrophy and disappearance of appendages.
Beneath the epidermis is a broad zone of pronounced lymphedema. Within this zone, the collagenous fibers are swollen and dilated, with numerous lymphatic channels filling the extracellular spaces. As the specimen undergoes dehydration, this process is enhanced, resulting in the formation of pseudobullae, which often are located intradermally.
Except in lesions of long duration, an inflammatory infiltrate is present in the dermis. The younger the lesion, the more prominent the inflammatory infiltrate. The mid portion and lower dermis may appear swollen, homogeneous, and eosinophilic, thus appearing sclerotic (hence *lichen sclerosus*).

Cases of overlap of morphea and LS may be seen and demonstrate the histologic changes of both disorders in their respective locations of the dermis.
Pathogenesis

Changes in the dermal matrix have been detected in LS. By electron microscopy, collagen fibrils often lack cross-striation, and in cross sections they sometimes have the appearance of empty tubes, suggesting degeneration of collagen. Fibrin and fibrinogen have been demonstrated in scleroderma and morphea, suggesting that this may be a nonspecific change.
The frequent finding of human papillomavirus alterations in lesions of LS have lead some to consider LS changes in cases of other chronic vulvar dermatitides. It has been hypothesized that persistent antigen may lead to the presence of CD8/CD57-positive T cells, which may play a role in tissue remodeling and fibrosis through cytokine elaboration.
In the epidermis, intercellular edema separates epidermal cells that show degenerative changes. There

Differential Diagnosis  .
Very early lesions may resemble lichen planus because of the apposition of the inflammatory infiltrate to the basal... and a subepidermal zone of edema usually has already begun to form in some areas in lichen sclerosus et atrophicus.

Old lesions of lichen sclerosus et atrophicus with thickening and eosinophilia of the collagen bundles in the...
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