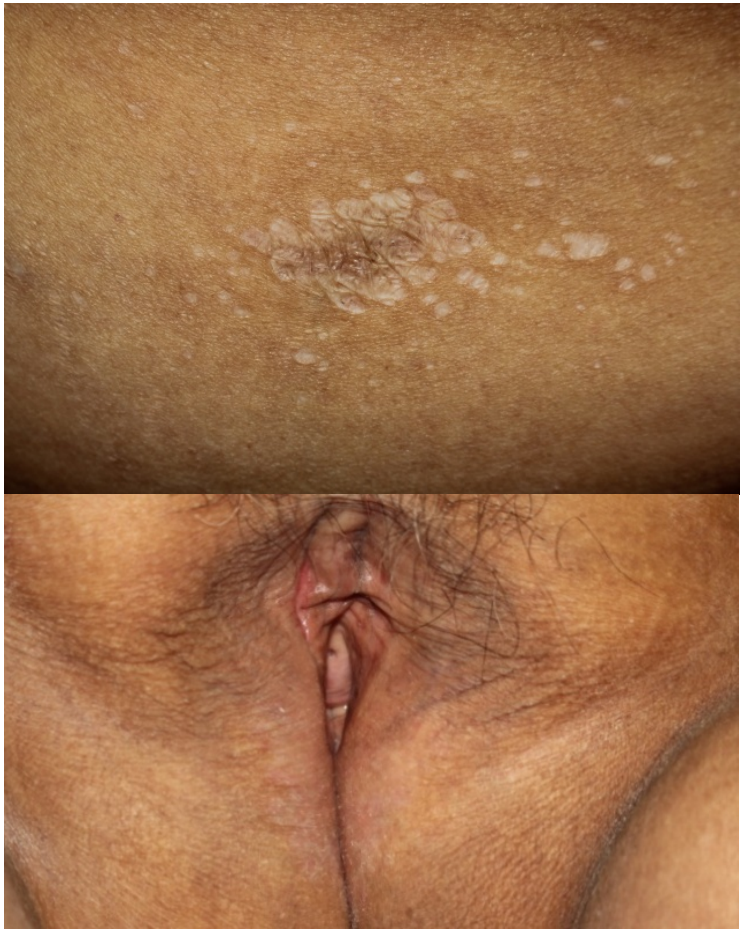
Case 20
A 61 year-old woman from Lopburi Province
Chief complaint: Multiple hypopigment macules and patches on trunk for 6 months.



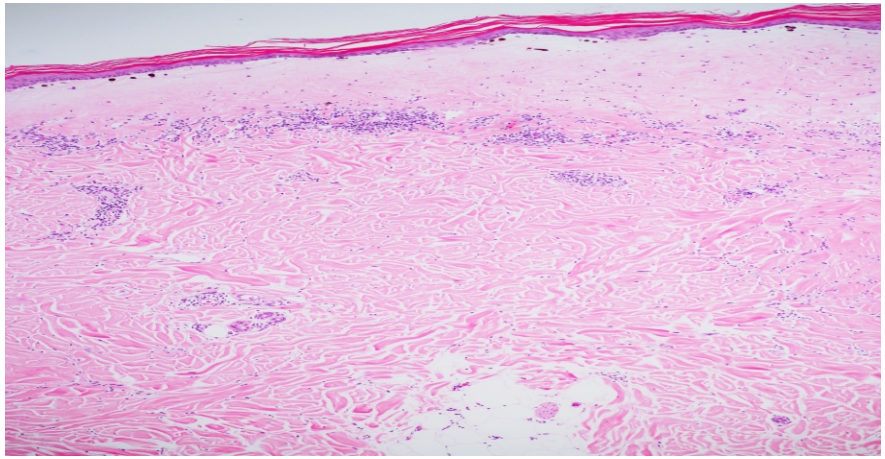
Present illness : The patient presented with 2-year history of itchy hypopigment patches on labia majora, perianal area and buttock, and 6 months PTA, she also developed symptomatic hypopigment macules and patches with wrinkled surface on abdominal wall. She applied unknown topical medication from pharmacy shop on the abdominal wall lesions, but her clinical did not improve.

Past history: Breast nodule BIRAD2 for 3 years
Family history: There was no family history of similar skin lesions and genitalia lesions.

Skin examination: Ill-defined hypopigment atrophic patches on right labia majora, buttock and perianal area, with superficial erosion on right labia majora. Multiple well-defined porcelain-white atrophic macules and patches on abdominal wall.

Physical examination: Systemic examination other than skin and genitalia lesions revealed no abnormality.

Histopathology: (S16-19869A, abdominal wall)



- Hyperkeratosis, epidermal atrophy and vacuolar alteration of basal cell layer
- Thickened homogenized collagen bundles with some melanophages, in the papillary dermis
- Perivascular and interstitial infiltrate of lymphocytes and a few plasma cell in the upper dermis

Diagnosis: Lichen sclerosus

Treatment: 0.05%Clobetasol propionate ointment apply twice daily.

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Discussion:

Lichen sclerosus (LS) is a chronic inflammatory skin disease that presents in anogenital and extragenital regions. LS is typically found in the anogenital region. Extragenital LS is less common, the incidence of extragenital LS is around6-20%.[1] LS occurs at all ages and both sexes. The male to female ratio varies between1:3 to 1:10.[2] It is more prevalent in females in the fifth or sixth decade and children younger than ten years.

Cause of LS is unknown. Presumably, there is a genetic predisposition. Approximately 10% of patients with LS have relatives with the same disease.[3] Immunological changes on the level of T and B cell have been described. Thus, an autoimmune phenotype has been observed in the case of vulvar lichen sclerosus involving increased level of Th1-specific cytokines, dense T cell infiltration and enhanced BIC/miR-155 expression as well as autoantibodies against extracellular matrix protein1 and BP180 antigen.[4-6] Oxidative DNA damage and TP53 mutations (tumor suppressor gene) have also been described. This could indicate an autoimmune background of lichen sclerosus and play a role in the slightly increased risk of vulvar carcinoma.[7]

LS can present on both anogenital and extragenital regions. Anogenital LS presents polygonal papules and porcelain-white plaques with atrophic fragile skin, fissures, telangiectasias, purpura, erythema, erosions and different degrees of sclerosis. Anogenital lesion causes severe discomfort, dyspareunia, dysuria, sexual dysfunction, genital bleeding or painful defecation. Progressive disease may lead to destructive scarring of genital organ. In female, LS could cause fusion of the labia minora and clitoris, narrowing of the vaginal introitus. In male, the lesion usually occurs on gland penis, prepuce, and foreskin remnants and may cause phimosis, painful erection, urethral stenosis may lead to urinary obstruction. Anogenital LS is associated with an increased risk 4-5% of malignant transformation[1-2,7-8], especially valvar squamous cell carcinoma. Thus the development of indurated plaque, nodule or ulcer arising LS indicate malignant transformation. Prompt clarification by means of a biopsy is imperative.[1,10] Extragenital LS usually presents with asymptomatic, whitish papules and plaques with wrinkled surface, typically affect the lip, neck, trunk and sites of trauma. Extragenital LS is not associated with malignant transformation.

Histopathology of classic lichen sclerosus, epidermis shows compact hyperorthokeratosis, atophy of the epidermis, follicular plugging, basal layer degenerative of keratinocyte, lichenoid infiltration at dermal-epidermal junction. In dermis shows subepidermal edema, absent elastic fiber, homogenization of collagen in upper dermis and lymphocyte infiltration. The differential diagnosis of LS and localized scleroderma (morphea) may be difficult in extragenital LS. Histologically of morphea, epidermis is relatively normal, no follicular plugging, no degeneration of basal layer. In palliary dermis shows homogenized collagen and elastic fiber and inflammation cell in dermis and

subcutis. The coexistence of morphea and LS can occur in the same biopsy specimen, the same location, or different location, usually genital LS.[10-12] The study recommended screening LS in morphea patient, especially anogenital inspection.[11]

Treatment of LS, the recommended initial treatment of lichen sclerosus is a three months application of potent to ultrapotent topical corticosteroids. Randomized studies show that application of potent to ultrapotent topical corticosteroids significantly improve LS in 75-90% of patients.[2] If the initial three months treatment does not lead to full remission in male patients with genital LS, a complete circumcision should be recommended.[13] Calcineurin inhibitor, topical tacrolimus and pimercrolimus are the second line treatment.[2] Using topical tacrolimus and pimercrolimus have shown clinical effectiveness in case series[13,14] and lesser side effect than steroids. Systemic therapy with retinoids, including isotretinoin, etretinate, acitretin[15] and oral tacrolimus have been useful in small trials. Phototherapy such as Narrow-band UVB[16], UVA1[17] and PUVA treatment have shown some improvement in LS. In asymptomatic extragenital LS require no treatment other than cover makeup in some cases. Prevent complication should be considered, especially in anogenital type significant complication including squamous cell carcinoma and corticosteroid adverse effects.

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