What’s Your Diagnosis?
Sharpen Your Physical Diagnostic Skills

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HISTORY
A 72-year-old woman with a history of hypertension and degenerative joint disease presents with severe vulvar pruritus of 10 years’ duration. The itching worsens with hot weather, anxiety, and stress. The patient also complains of intermittent dysuria. She feels depressed and remains isolated because of embarrassment related to the constant vulvar itching.

She underwent a subtotal hysterectomy with bilateral salpingo-oophorectomy in 1982 for menorrhagia, and she also had colon surgery in 1992 for intestinal perforation.

PHYSICAL EXAMINATION
Results of physical examination of the vulva are shown here. No inguinal lymphadenopathy is noted.

LABORATORY RESULTS
Laboratory results are unremarkable except for elevated total cholesterol. Papanicolaou smear from the cervix is negative for human papillomavirus and cervical intraepithelial neoplasia. Punch biopsy of the vulva is done, 4 mm of tissue taken from the right labium majus and 4 mm of tissue from the left labium majus, and the specimens are sent for histopathological examination, which confirms the diagnosis.

WHAT’S YOUR DIAGNOSIS?

Answer: Lichen sclerosus chronicus

Vulvar lichen sclerosus is a chronic inflammatory dermatosis. This progressive and mutilating disease often goes undetected for years. It is characterized by clinicopathologic persistence and hypocellular fibrosis (sclerosis). Advanced disease severely affects quality of life and even may cause psychological side effects. This condition was previously designated as lichen sclerosus et atrophicus; this term was dropped because areas of thickening and hyperkeratosis often occur as in this patient. Any skin surface can be affected, but 85% to 98% of cases occur in the anogenital region.

ETOLOGY
It is one of the most common conditions treated in vulvar clinics. Multiple causes have been proposed, including genetic, immunological, and local skin factors, but hormonal factors are highly correlated with pruritus vulvae in lichen sclerosus. Research studies show that it is common in premenarchal children and postmenopausal women with estrogen deficiency.

CLINICAL FEATURES
Vulvar pruritus is the hallmark of this disease; other features are soreness or irritation, dyspareunia and dysuria due to fissuring, introital stenosis, and fusion of labia minora. Classically, lichen sclerosus is expressed as white atrophic papules that coalesce together, but it can also be hemorrhagic, purpuric, hyperkeratotic, or ulcerated.

Most lesions affect the labia minora and/or majora, although they may extend and surround the anus in a keyhole fashion. Thickening is due to scratching, which may result in excoriations and lichenification. The main diagnostic tool is skin biopsy; it confirmed the diagnosis of lichen sclerosus in this patient.

ASSOCIATION WITH SQUAMOUS CELL CARCINOMA
Vulvar lichen sclerosus could act as initiator and promoter of squamous cell carcinoma (SCC), explaining the frequent coexistence of these two pathologies. The
histological changes of lichen sclerosus are often associated with vulvar SCC due to chronic inflammation and scarring, which are strongly related to oncogenesis; this association has been well documented. A subset of vulvar SCCs associated with lichen sclerosus diffusely express the tumor suppressor gene p53 and may be involved early in the process of carcinogenesis.

TREATMENT

The primary medical therapy consists of superpotent topical corticosteroid ointments. Early aggressive topical corticosteroid therapy also relieves symptoms of urethral strictures.

Betamethasone dipropionate 0.05% ointment was given to this patient for topical use for 1 week. The Figure shows the appearance of the lesion after corticosteroid treatment. The patient’s symptoms completely resolved.

Women with estrogen deficiency can be given hormonal treatment, but it does not improve symptoms. Other treatment modalities are immune modulators, systemic retinoids, progesterone, cryosurgery, carbon dioxide laser, and vulvectomY, but these have high recurrence rates.

SCREENING

Vulvar disease is uncommon, and there is no evidence that screening would decrease the incidence.

DIFFERENTIAL DIAGNOSIS

Lichen planus. This relatively uncommon inflammatory dermatologic condition affects the skin, nails, and mucous membranes. The peak age of onset is 50 to 60 years. It usually presents with intense pruritus or vulvar pain, soreness, or burning and vaginal discharge. Vaginal discharge that does not respond to treatment is a prominent symptom in lichen planus but is not found in lichen sclerosus chronicus. Grossly, lesions are characterized by glassy, brightly erythematous erosions with striae (Wickham’s striae) along the margins. Occasionally, a violaceous border is noted as well.

Vitiligo. This acquired skin pigmentation disorder is more disfiguring in blacks. Genetic causes are found in 20% to 30% of cases. It is thought that an autoimmune process directed against melanocytes is involved. Pruritus is a frequent symptom. Diagnosis is based on vulvar biopsy. Microscopically, there is evidence of squamous differentiation with keratin deposits and intercellular bridging.

Paget’s disease of vulva. This very rare disease has a high misdiagnosis rate. The significance of the disease lies in its association with underlying malignancy. It mainly affects the epidermis and occasionally extends to the underlying dermis. It has a predilection for apocrine gland–bearing areas, axilla, perineum, and vulva. The management is notoriously complicated, and recurrence rates are high despite aggressive surgeries.

references: