Series Editor: William W. Huang, MD, MPH

Vulvar Diseases, Part 2

Rita Pichardo-Geisinger, MD

Dr. Pichardo-Geisinger is Associate Professor of Dermatology, Wake Forest Baptist Medical Center, Winston-Salem, North Carolina. The author reports no conflict of interest.

Disease	Symptoms	Clinical Features	Associations	Management	Other
Behçet disease	Painful genital ulcers	Oral and genital aphthous ulcers; skin lesions present as sterile papulopustules and palpable purpura to erythema nodosum	Recurrent oral ulceration, recurrent genital ulceration, arthritis, ocular abnormalities (eg, uveitis, retinal vasculitis)	Azathioprine, cyclophosphamide, cyclosporine, methotrexate, TNF-α inhibitors, etanercept, infliximab, thalidomide	The presence of any 2 or more of the following indicates a diagnosis of Behçet disease: genital aphthosis, skin lesions, eye lesions, and positive pathergy test
Crohn disease	Suprapubic pain	Edema on the vulva, perianal skin tags, abscesses and/or fistulas, linear "knife-cut" ulcers in skin folds	Cutaneous lesions, joint involvement, ophthalmic disease	Systemic corticosteroids, methotrexate, sulfasalazine, or metronidazole either individually or in combination; intralesional triamcinolone injections (10–20 mg/mL)	Hidradenitis suppurativa with anogenital lesions sometimes is confused with Crohn disease; mutations in the *NOD2, ATG16L1,* and *IRGM* genes are present
Extramammary Paget disease	Pruritus is the initial symptom; burning; can be asymptomatic	Red plaques with a rough surface with erosions or white thickened islands on a keratinized genital epithelium	Urothelial carcinoma	Excision via Mohs surgery (>1 mm invasion justifies evaluation of lymph nodes); laser surgery, radiation therapy, topical fluorouracil, imiquimod	Reported more often in individuals aged >50 y; women are more often affected than men; 10%–20% of patients have underlying visceral malignancy
Pemphigus vulgaris	Painful erosions	90% of patients have mucosal involvement; erosions on the labia minora and majora, vagina, and cervix	Pemphigus vegetans is a variant that presents with erosions and peripheral pustules in early phases and vegetating and verrucous plaques in later phases	Mainstay of therapy is oral prednisone (60–150 mg/day) as well as steroid-sparing immunosuppressive agents (eg, azathioprine, cyclophosphamide); dapsone, antimalarials, cyclosporine, plasmapheresis and IVIG, mycophenolate mofetil, rituximab	Severely affected vulva exhibits resorption of the labia majora and clitoris with scarring

continued on next page

(continued)

Disease	Symptoms	Clinical Features	Associations	Management	Other
Perianal streptococcal dermatitis	Persistent pruritus or pain	Erythema, fissuring, fragility, crusting, exudation and erosion of the perianal skin; group A β-hemolytic streptococcus is the causative organism	Mucopurulent vaginal discharge, guttate psoriasis	Oral penicillin with topical mupirocin several times daily	Skin should be cultured to confirm diagnosis; recurrence is common; affects children
Plasma cell vulvitis	Pruritus, soreness, and burning; can be asymptomatic	Usually presents as a solitary, well-demarcated, deep to rusty red in the vestibule	Lichen planus	Potent topical corticosteroids, intralesional corticosteroids, imiquimod, tacrolimus, pimecrolimus, CO_2 laser	Differential diagnosis includes lichen planus, inverse psoriasis, candidiasis, VIN; equivalent to plasma cell balanitis in males
Vulvodynia	Burning, stinging, irritation, aching, soreness or throbbing	Absence of objective findings of skin disease; pain to touch with a cotton-tipped applicator limited to or worst in the vestibule	Other chronic pain syndromes (eg, headaches, fibromyalgia, irritable bowel syndrome, interstitial cystitis, temporomandibular joint syndrome), sexual dysfunction, depression and anxiety	Pelvic floor physical therapy, treatment for depression and/or anxiety (eg, counseling, sex therapy, oral tricyclic antidepressants), oral therapy for neuropathic pain (gabapentin, venlafaxine, tricyclic antidepressants, duloxetine), vestibulectomy, botulinum toxin A	Pelvic floor muscle abnormalities have been recognized as a causative factor

Abbreviations: TNF-α, tumor necrosis factor α; *NOD2*, nucleotide-binding oligomerization domain containing 2; *ATG16L1*, autophagy related 16-like 1; *IRGM*, immunity-related GTPase family M; IVIG, intravenous immunoglobulin; CO_2, carbon dioxide; VIN, vulvar intraepithelial neoplasia.

Practice Questions

1. **A 5-year-old girl presented to your clinic with an itchy rash in the vulvar and anal regions. The patient's mother reported erythema and erosion of the anal area. Her pediatrician prescribed an oral antibiotic that showed good results but the condition recurred 2 weeks after she finished the medication. The most likely diagnosis is:**
   a. Behçet disease
   b. pemphigus vulgaris
   c. perianal streptococcal dermatitis
   d. plasma cell vulvitis
   e. vulvodynia

2. **A 34-year-old woman presented with pain and a burning sensation on the vulva. She reported a history of migraines. On physical examination, mild erythema was noted on the labia majora and minora and the patient reported pain to the touch of a cotton-tipped applicator in the vestibule. The most likely diagnosis is:**
   a. Crohn disease
   b. extramammary Paget disease
   c. pemphigus vulgaris
   d. plasma cell vulvitis
   e. vulvodynia

3. **A 25-year-old woman with a history of oral ulcers presented to your clinic with pain in the genital area. On physical examination, multiple ulcers were noted on the labia majora with no discharge. The most likely diagnosis is:**
   a. Behçet disease
   b. Crohn disease
   c. extramammary Paget disease
   d. pemphigus vulgaris
   e. plasma cell vulvitis

4. **A 56-year-old woman presented to your clinic with vulvar pruritus and a burning sensation of 6 months' duration. She had used a topical antibiotic and hydrocortisone cream 1% without relief. On physical examination, a red, irregular plaque is noted on the vestibule. The most likely diagnosis is:**
   a. Behçet disease
   b. extramammary Paget disease
   c. pemphigus vulgaris
   d. plasma cell vulvitis
   e. vulvodynia

5. **A 44-year-old woman presented to your clinic with pain and edema of the vulva. At physical examination, erythema and fissures were noted around the anus with fistulas involving the perianal skin. What is the most likely diagnosis?**
   a. Behçet disease
   b. Crohn disease
   c. extramammary Paget disease
   d. pemphigus vulgaris
   e. plasma cell vulvitis

Fact sheets and practice questions will be posted monthly. Answers are posted separately on www.cutis.com.