Hidradenitis Suppurativa

Background

1. Definition:
    - Chronic, recurrent skin disease with occlusion and inflammation of hair follicles affecting intertriginous skin of axillary, inguinal and anogenital areas
    - Lesions may present as painful nodules, bands of severe scarring and draining sinuses, eventually leading to fistula formation
2. General information:
    - Hidradenitis suppurativa (HS) marked by periods of inflammation with occasional 2° infection, and intermittent remission that can last several years
    - Exacerbations precipitated by sweat, heat, stress, tight clothes and friction from adiposity in intertriginous areas

Pathophysiology

1. Pathology of disease:
    - Etiology and pathogenesis not clear
    - Proposed mechanism by several authors including Yu, Boer, and Heller[2,3]
        - Occlusion of hair follicle with follicular plugging
        - Occlusive spongiform infundibulofolliculitis weakens infundibular wall → duct rupture
        - Inflammatory reaction to rupture → spread to adjacent apocrine and eccrine glands? further tissue destruction and skin damage
        - Bacterial infection secondarily
2. Incidence, prevalence:
    - Incidence of HS: 1 in 300, affecting races equally
    - Female: Male ratio 3:1
    - Onset of symptoms: puberty to age 40[4]
    - Avg. duration of single lesion: 7 days
    - Avg. number of lesions: 2/month (range: 1/year – 30/month)
    - Many patients present with at least 1 active lesion at all times[5]
3. Risk factors:
    - Overweight/Obesity – Shearing vs. hormonal effects[6]
    - Endocrine:
        - HS rarely seen before puberty
        - Childhood cases of HS associated with precocious puberty[7]
        - Diabetes, Cushing's disease, acromegaly correlated[8,9]
    - Genetic Factors:
        - Familial predisposition, possibly genetic in origin, with autosomal dominant and heterogeneous patterns noted[10,11]
    - Medications:
        - Lithium, and oral contraceptives with medroxyprogesterone acetate have been reported as potentially etiologic agents[12,13]
    - Associated Conditions:
        - Acanthosis nigricans
        - Arthritis (certain forms)[14]
        - Crohn's disease
        - Down syndrome

        - Graves' disease
        - Hashimoto's thyroiditis
        - Herpes simplex
        - Hyperandrogenism
        - Irritable bowel syndrome
        - Sjogren's syndrome
    4. Morbidity / mortality:
        - Cellulitis
        - Systemic infection
        - Fistulae
        - Arthropathy (2° to inflammatory injury)
        - Depression
        - Lymphedema (2° to lymphatic obstruction)
        - Anemia (2° to chronic infection)
        - Hypoproteinemia (2° to chronic infection/drainage)
        - Amyloidosis (2° to chronic infection)
        - Squamous Cell Carcinoma (in indolent sinus tracts)[15]

Diagnostics

1. History:
    - Discomfort, itching, erythema, burning, and hyperhidrosis in commonly affected areas
    - May include tender or painful nodule +/- malodorous drainage
    - Fever and sepsis rare
2. Physical examination:
    - Tender, firm, painful nodule(s) in apocrine rich areas: axillae, periareolar, intermammary, pubic, infraumbilical, gluteal, inguinal, and perianal regions with or without signs of cellulitis
    - Sinus tracts, draining fistulas may mark chronic dx or fibrous banding/scarring
3. Diagnostic testing
    - Usually a clinical diagnosis
    - Culture of drainage may help guide antibiotic treatment
    - Biopsy – especially in cases of perianal hidradenitis to exclude coexisting cancer or Crohn's disease[15]
    - TSH (due to association of Hashimoto's)
    - If patient is febrile or toxic consider CBC/blood culture
4. Diagnostic criteria:
    - **Hurley's Clinical Staging**:[16]
        - **Stage I:**
            - Abscess formation without sinus tracts
            - Can be managed with drug therapy
        - **Stage II:**
            - Recurrent abscesses with tract formation
            - Managed with drug therapy and limited excision of recalcitrant lesions

- **Stage III:**
  - Diffuse or near diffuse involvement, or multiple interconnected tracts and abscesses across the entire area
  - Surgical treatment/excision indicated

Differential Diagnosis

1. Acne
2. Crohn's dz
3. Epidermal Inclusion cyst
4. Furuncle
5. Follicular pyodermas
6. Granuloma inguinale
7. Lymphogranuloma venereum

Therapeutics

1. Acute treatment:
   - There is no cure but treatment options are available
   - NSAIDs (for pain management, reduction of inflammation)
   - Hot compresses, topical antibacterials, weight loss, smoking cessation
   - Antibiotics:
     - Systemic therapy if cellulitis
     - Antistaph agents best for axillae
     - More broad spectrum coverage best for perianal
     - Clindamycin (300 mg PO BID) (SOR:2B)[4,18]
     - Rifampin (300 mg PO BID for 12 weeks) (SOR:2B)[4,19]
     - Dicloxacillin (1-2 g PO daily) (SOR:3C)[4,19]
     - Erythromycin (1 g PO daily) (SOR:3C)[4,19]
     - Minocycline (1 g PO daily) (SOR:3C)[4,19]
     - Tetracycline (500 mg PO BID)[18]
   - Topical treatments:
     - Clindamycin (1%) (SOR:2B)[4]
   - Incision and drainage if abscess (SOR:2C)[4]
2. Further management (mostly for chronic dz)
   - OCP
     - High estrogen: progesterone ratio
     - Low androgenicity of progesterone
   - Oral retinoids: isotretinoin
   - Corticosteroids (systemic, intralesional and topical)
   - Immunosuppressants:
     - Cyclosporine (limited studies, but benefit shown)[20]
   - Dapsone 25-150 mg PO daily (SOR:3C)[4]
   - Surgery:
     - I&D for acute dz
     - More extensive options for chronic disease
   - Other treatments:
     - C02 laser: (SOR:3C)[4,21]
     - Photodynamic therapy: seems temporary aide[22,23]
     - Cryotherapy (SOR:3C)[4]
       - Total doses 3-8 Gy resulted in great improvement of symptoms[4,17]

3. Long-term care
    - Severe, recurrent dz:
        - Anecdotal evidence suggests 2 months or more of antibiotic treatment prevents progression and worsening of concomitant infection[4]
    - Finasteride
        - Case series by Joseph et al showed an improvement of HS in both male and female patients with the use of Finasteride[24]
    - Anti -TNF alpha immunomodulators:
        - Infliximab
        - Many studies have been conducted demonstrating improvement in patients with Crohn's and HS
        - Other studies have shown an improvement in symptoms even in those patients not afflicted by Crohn's dz)[25,26]
    - Consider referral to dermatologist if other treatment options not successful
    - Consider referral to general surgeon especially in severe, disfiguring disorder

Follow-Up

1. Return to office
    - As a chronic disease scheduled follow-up suggested
    - Return to clinic sooner if pain is persistent, failure of antibiotics, or conditions worsen
2. Refer to specialist
    - Dermatology, surgery, psychology as needed
3. Admit to hospital
    - If septic or resistant infection

Prognosis

1. Nodules will heal slowly, with or without drainage, over 10-30 days
2. In typical cases, nodules recur at least several times yearly
3. In severe cases, the patient may suffer a constant succession of new lesions forming as soon as old lesions heal
4. Remissions may last from months, or years

Prevention

1. Avoid deodorants, and shaving if these cause irritation
2. Warm compresses, topical antiseptics, antibacterial soap aid with folliculitis
3. Weight loss will decrease adiposity, and reduce friction in intertriginous areas
4. Avoid prolonged exposure to hot, humid climates

Patient Education

1. Hidradenitis Suppurativa: What You Should Know
   http://www.aafp.org/afp/20051015/1554ph.html

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