Atypical Wounds

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Chronic Wounds

- Incidence of chronic wound in US is 6 million per year
- Majority ????
- 10% of lower extremity ulcers are due to less frequent etiologies
  - Inflammatory processes
  - Infection
  - Metabolic disorders
  - Neoplasms

Pyoderma Gangrenosum (PG)

- Non-infectious, progressive necrotizing skin disorder
- Etiology unclear
- Diagnosis of exclusion

Prevalence and Incidence

- Occurs 1 in 100,000 of population
- Age 20-50
- Predominantly females
- 50% have other systemic dx such as inflammatory and/or GI disorders
- 30% have pathergy

Signs and Symptoms

- c/o pain – stabbing or out of proportion with wound characteristics
- Ulcer location typically on Lower Extremities, sometimes trunk
- Ulcers begin as nodule, blister or pustule
- Borders – raised, irregular, sharp, margined, undermining, purple or gray
- Rapid progression that spread and increase significantly in size within days with increased necrosis to periwound and wound bed
- Can be recurrent

Diagnosis of Exclusion

- Document clinical presentation
- Order cultures, labs, biopsy and vascular studies
- Need to exclude “other” diagnosis
Differential Diagnoses

- Venous Leg Ulcer
- Vasculitis
- Trauma
- Drug reactions
- Bites
- Non-healing burn

Treatment Recommendations

- 1-2 mg/dg/day prednisone
- Pulsed IV 1g/day for 3-5 days if rapid treatment needed
- Low dose cyclosporin 3-5 mg/kg/day as primary or adjunct if corticosteroids fail
- Dapsone as maintenance therapy with or without prednisone
- Moisture retentive dressings for pain control, induce collagen production, facilitate autolytic debridement and promote angiogenesis
- Irrigation for bacterial and fungal growth
- Topical Triamcinolone Cream [TAC] to wound and borders twice weekly
- Surgical or sharps debridement contraindicated

Case Study

- 58 yo African American female with recurrent ulcers to bilateral lower extremities
- Recent treatment in Wound Center 2 years prior to this admission for venous insufficiency
- 7 year Hx of Diabetes
- Hx of recurrent ulcers to lower extremities
- HTN
- Asthma
- Recent cough
- Denied ulcerative colitis, Crohn’s disease, Inflammatory bowel related disease

Initial Visit

- (May) – 9 full thickness wounds with purple or lavender borders, moderate slough, minimal granulation, good extremity pulses, complaints of pain (burning, stinging, tingling)
- Initial Dx - DWLE Grade 1 with underlying venous disease and possible Pyoderma Gangrenosum
- Work Up:
  - labs for infection, inflammation, nutritional status, baseline kidney and liver function
  - Ultrasounds to rule out arterial and venous disease – no insurance
  - Cultures of wounds
  - Biopsy of wound
- Treatment – Selective sharps debridement and dressed with cadexamer iodine and light compression to be changed two times weekly

Follow Up Visit

- Biopsy showed acute neutrophilic inflammation with necrosis and ulceration consistent with Pyoderma Gangrenosum
- Cultures – 3+ acinetobacter, 3+ strep, 3+ corynebacterium treated with augmentin
- Labs – elevated glucose, low prealbumin, elevated ESR
- No change in wounds except two new wounds
- POC – Dx changed to PG
- Weekly MD for selective debridement
- Nursing 2-3 x weekly for dressing changes

Progressive Plan of Care

- Over the course of 6 months:
  - Several antimicrobial dressings including silver, cadexamer iodine, antibiotic ointment, methylene blue and gentian violet
  - Monthly cultures requiring several rounds of antibiotics including for MRSA
Progressive Plan of Care

For first three months:

- Wounds would improve then deteriorate
- Patient had negative reactions to some topical agents

Progressive Plan of Care

Fourth month:

- Developed rash – discontinued current antibiotics, initiated topical silver and ordered Benadryl
- Next week – 3 wounds had healed

Progressive Plan of Care

Fifth month:

- Steadily healed 2-3 more wounds each week
- Zyvox initiated in October for MRSA
- Approved for Indigent Care by treating facility
- Referred to Infectious Disease – initiated 10mg Prednisone TID

Progressive Plan of Care

Sixth month:

- Initiated Triamcinolone Cream (TAC) in November to wounds
- Dec 2 – all wounds completely healed
- Referred again for ultrasounds to check arterial and venous insufficiency – negative for both

Outcome

- PG effectively suspected, excluded and included
- Likely misdiagnosed two years prior
- Did not treat with steroids initially
- Due to pain, irritation and rash, dressings sometimes changed weekly to something different making tracking progress difficult
- Wound began to improve prior to steroid initiation
- Selective debridement contraindicated for PG
- Lack of insurance was an obstacle
- Outcome ultimately achieved healing but could wounds have been healed quicker????

Hydradenitis Suppurativa

- Considered a severe form of acne occurring deep around the sebaceous glands and hair follicles
- Chronic skin inflammation with blackheads and/or bumps/lesions that break open and drain pus
- Groin and armpits where apocrine sweat glands are located
- Generally appears after puberty

http://www.mayoclinic.com/health/hidradenitis-suppurativa/DS00818
Prevalence and Incidence

• 1-2% of general population
• All races but increased in African Americans
• Seen greater in hot, humid environments
• More women than men
• Men – greater in anogenital region
• Females - greater in axilla
• Onset anytime between puberty and post menopause – ages 11-50

Risk Factors

• Obesity
• Smoking
• Family history of acne
• Apocrine duct obstruction
• Secondary bacterial infection
• Hirsutism
• Chemical irritants – deodorants or antiperspirants
• Mechanical irritants – shaving or depilatory use

Signs and Symptoms

Early:
• Itching
• Erythema
• Excessive localized perspiration

Late:
• Lesions
• Pain
• Purulence
• Disfigurement

Signs and Symptoms

• Papules or nodules
• Abscesses
• Inflamed
• Erythema
• Purulent
• Dermal contractures and ropelike elevation of the skin
• Double-ended (bridged) comedones

Diagnosis

• Clinical findings
  • Characteristic lesions
  • Typical distribution of lesions
  • Recurrence – remissions of long periods may delay diagnosis
• Must have one of the following:
  • One active primary lesion and history of 3 or more discharging and painful lesions since puberty
  • Inactive disease (no current lesion) but history of 5 or more painful and draining abscesses since puberty
• Labs
  • CBC with diff, ESR, CRP, CMP, urinalysis, consider thyroid and anemia workup
• Cultures
  • Ensures appropriate antibiotics
  • Usually grow staph and/or strep

Differential Diagnosis

• Associated comorbidities:
  • Crohn’s Disease
  • Irritable Bowel
  • Certain arthritis
  • Down Syndrome
  • Graves Disease

• Mimics
  • Folliculitis
  • Furunculosis
  • Pilarial cysts
  • Actinomycosis
  • Cat scratch disease
Treatments

- Local hygiene – soaps without dyes and perfumes
- Weight reduction
- Warm compresses
- Loose fitting clothes
- Absorptive Antimicrobial and/or charcoal dressings for odor
- Oral Antibiotics – to reduce inflammation – abx used for acne (erythromycin, tetracycline, minocycline, doxycycline)

Treatments (continued)

- Corticosteroid injections into around lesions
- NSAIDS to manage pain
- I&D if large and fluctuant or painful nodules
- Radical surgery (aggressive approach) but very effective if late stages – must remove the entire affected and scarred area
- NPWT and/or skin grafting if surgical option chosen
- Specialist Referrals – Infectious Disease, Plastic Surgeon, Surgeon, Immunologist
- HBO – not CMS or UHMS approved
  - Resources: www.familydoctor.org; www.hs-usa.org; www.aafp.org

Vasculitis

- Autoimmune disease causing inflammatory changes in blood vessels leading occlusion causing poor lumen integrity, bleeding, ischemia and necrosis
- Rare, chronic and relapsing disease
- Can affect large and small vessels

Prevalence/Incidence

- Men > Women
- Onset ages 65-74 yo

Risk Factors

- Autoimmune disorders
  - RA
  - SLE
  - Sjogren’s Syndrome

Signs and Symptoms

- Deep, punched out ulcers
- Red, purple or blue wound edges
- Painful
- Rapid deterioration
- Purpuric rash
Diagnosis

- Clinical findings
  - Patient History
  - Wound Appearance

- Labs
  - ESR
  - CRP
  - Platelets
  - WBC
  - Thrombolytic panel

- Biopsy
  - Perilesional skin
  - R/O Malignancy

- Culture
  - Infection

- Immunological tests
  - Rheumatoid Factor (RF)
  - Anti-nuclear Antibody (ANA)
  - Low Serum Complement
  - Antineutrophil Cytoplasmic Antibodies (ANCA)

Differential Diagnosis

- Thrombolytic Disease
- Embolic Disease

Risk Factors

- History of smoking
- Onset before age 50
- Upper and/or Lower Extremity vessel involvement without atherosclerosis or common risk factors
- Popliteal arterial occlusions

- Inclusion Criteria to Diagnose – must have all above EXCEPT upper extremity involvement

Buerger Disease (Thromboangiitis Obliterans)

- “Nonatherosclerotic vaso-occlusive inflammatory disease” of the small and medium distal arteries
- Etiology or cause is unknown
- Primary association with tobacco use

http://www.hopkinsvasculitis.org/types-vasculitis/buergers-disease/

Treatments

- Systemic Treatment of causative factors
  - Steroids
  - Anti-inflammatory
  - Antihistamines
  - Immunosuppressants

- Pain control
- Topical wound care
- Multidisciplinary communication

Diagnosis

- Inclusion Criteria to Diagnose – must have all EXCEPT upper extremity involvement

- Labs
  - Exclude collagen vascular disease
  - Exclude hypercoagulable state
  - Exclude high cholesterol

- Radiographic Imaging
  - Exclude arterial calcification
Signs and Symptoms

- Claudication
- Pain in distal extremities at rest
- Painful ulcers in extremities
- Limb amputations frequent

Differential Diagnosis

- Raynaud’s Phenomenon
- Vasculitis
- Arteriosclerotic “Arterial” Disease
- Frostbite

Treatments

- Smoking Cessation – must stop to prevent progression
- Pressure Redistribution
- Topical agents for wound healing
- HBO – not CMS or UHMS approved
- Surgical debridement
- NPWT
- Skin grafting

Calciphylaxis

- Vessel calcification with thrombosis and skin necrosis
- Rare and serious disease
- Primarily seen in patients with ESRD
- Calcific Uremic Arteriolopathy

Prevalence/Incidence

- 1% incidence per year
- 4% prevalence in patients with ESRD
- Prognosis – poor
- 63% mortality if proximal skin lesions
- 23% mortality if distal skin lesions
- 39% mortality within 6 months of being diagnosed
- Mortality increases to 80% if skin ulcers develop

Risk Factors

- End Stage Renal Disease
  - Diabetes
  - Peritoneal Dialysis
  - Hypoalbuminemia with chronic inflammation
  - Malnutrition
  - Hypertension
  - Atherosclerosis
  - Hyperphosphatemia
  - Hypercalciemic states
  - Milk-alkali syndrome
  - Hypervitaminosis D
  - Elevated calcium-phosphate product
**Signs and Symptoms**
- Painful red to purple livedoid plaques
- Reticulated, violaceous and mottled patches
- Rapid progression to non-healing necrotic ulcers
- Vesicles at periphery
- Bullae
- Eschar or gangrene
- Subq nodules extending centimeters from edge of lesions

**Diagnosis**
- Clinical Findings
  - Patient history
  - Wound appearance
- Biopsy
  - Trauma?
  - Incisional cutaneous bx preferred
  - Looking for small vessel calcification with endovascular fibrosis, panniculitis, tissue necrosis

**Diagnosis**
- Clinical Findings
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**Differential Diagnosis**
- Diabetic Wound of the Lower Extremity
- Gangrene
- Arterial Wound
- Pressure Ulcer

**Treatments**
- No evidence based guidelines
- Prevention
- Systemic treatment
  - Increase dialysis frequency
  - Adjustments in procedures
- Partial parathyroidectomy
- Wound Care
  - Debridement – aggressive vs conservative due to pain
  - HBO – no RCTs and not an approved CMS/UHMS diagnosis
  - Topical wound care based on ulcer characteristics

**Epidermolysis Bullosa (EB)**
- Genetic blistering skin disease with fragile skin leading to bulla formation
- Multiple forms
- Mucosa involved

[Image: http://www.dermnet.com/images/Epidermolysis-Bullosa/]
**Types of EB**

- **EB Simplex** – most common
  - Blisters at basal layer of epidermis
  - Soles and palms
  - Aggravated with heat and friction

- **Junctional EB**
  - Blisters at lamina lucida level of basement membrane
  - Periorificial areas, ocular, tracheolaryngeal, GI, GU, renal

- **Dystrophic EB**
  - Blisters beneath lamina densa within dermis
  - Areas prone to “knocks”
  - Pseudosyndactyly in more debilitating cases

**Treatment**

- Minimize trauma
- Blisters intact
- Wound care of open blisters
- Immunosuppressants
- Anti-inflammatory
- Address pain, itching and nutrition
- Consider EB specialized clinic (16 countries, April 2013)
- Resources: www.debra.org/international, www.internationalebforum.org

**Bullous Pemphigoid**

- Autoimmune blistering disease
- Typically seen in the elderly
- Pruritis common


**Prevalence/Incidence**

- Uncommon
- Frequency unknown
- Onset age 65-76 years
- Exacerbational Disease

**Risk Factors**

- UV Irradiation
- Xrays
- Drugs
- Vaccination in children

**Signs and Symptoms**

- Tense blisters with thick roofs
- Severe itching
- Multiple types:
  - Generalized Bullous Form – most common
  - Vesiculocyst Form – groups
  - Vegetative – plaques in axilla, neck groin, inframammary areas
  - Generalized erythodema – rare – resembles exfoliative skin conditions (Psoriasis)
  - Urticarial – hive-like that progress to bullous
  - Nodular – rare
  - Acral – childhood onset with vaccination
  - Infant – acral or generalized
Diagnosis

- Biopsy
  - Edge of blister

- Gold standard
  - Immunofluorescence (IF) microscopy

Differential Diagnosis

- Epidermolysis Bullosa
- Bullous Diabeticorum

Treatments

- Topical Treatment
  - Reduce blister formation
  - Epithelialization of open areas
  - Topical Steroids over a wide area

- Systemic Treatment
  - Anti-inflammatories
  - Immunosuppressants
  - Prednisone
  - Initial dose not to exceed 0.75mg/kg/d
  - Reduce meds to control disease but reduce side effects
  - Treatment may take 6-60 months until remission
  - Exacerbation treatment

Summary

- LISTEN
  - Make sure to Include AND Exclude
  - Listen for recurrence or exacerbations
  - Don’t discount co-morbidities that you think are “unrelated” to the wound
  - Re-evaluate if no healing in 4 weeks
  - Think outside the box
  - Multidisciplinary approach

Questions???

References

References


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References