NOTES
URTICARIA & ERYTHEMA NODOSUM

GENERALLY, WHAT ARE THEY?

PATHOLOGY & CAUSES

- Vascular reaction of the skin triggered by allergic reaction, irritation, or infection
- Vasodilation, increased vascular permeability → fluid leaks into interstitium → swelling/edema
- Possible elicitation of hypersensitivity reaction (immune system involved)
- Can be acquired (e.g. medications), associated with underlying illness (e.g. malignancies, autoimmune disorders), or have genetic predisposition

SIGNS & SYMPTOMS

- Range of dermatological manifestations:
  - Erythema
  - Swelling
  - Urticaria, pruritus
  - Raised or flat lesions

DIAGNOSIS

- Physical examination
  - Based on appearance
- Patch testing to confirm and determine the allergy
- Screening for autoimmune or neoplastic etiologies

TREATMENT

- Identify/avoid triggers
- Address underlying cause
- Symptomatic management

ERYTHEMA NODOSUM

osms.it/erythema-nodosum

PATHOLOGY & CAUSES

- Acute skin eruption due to inflammation in the subcutaneous adipose tissue
  - Most common form of acute panniculitis
- Chronic or recurrent forms are rare but may occur
- Presumably caused by a delayed hypersensitivity type IV reaction to a variety of antigens

CAUSES

- 30–50% unknown etiology
- *Infections*: *Streptococcus spp.*, *M. tuberculosis complex*, *M. leprae*, *M. pneumoniae*, *Yersinia spp.*, *Histoplasma capsulatum*, *Coccidioides immitis*
- *Autoimmune disorders*: inflammatory bowel disease, sarcoidosis, Behçet's disease, medium-vessel vasculitis
- *Medications*: sulfonamides, oral contraceptives, amiodarone

- *Malignancies*: hematological malignancies, carcinoid tumours, pancreatic cancer

SIGNS & SYMPTOMS

- Pre-eruptive phase
  - Fever, malaise, and arthralgia
- Eruptions of red, painful, poorly defined plaques and nodules, most commonly located on shins, knees, arms, thighs, and torso → skin lesions gradually get softer and smaller until they completely disappear over the course of about two weeks

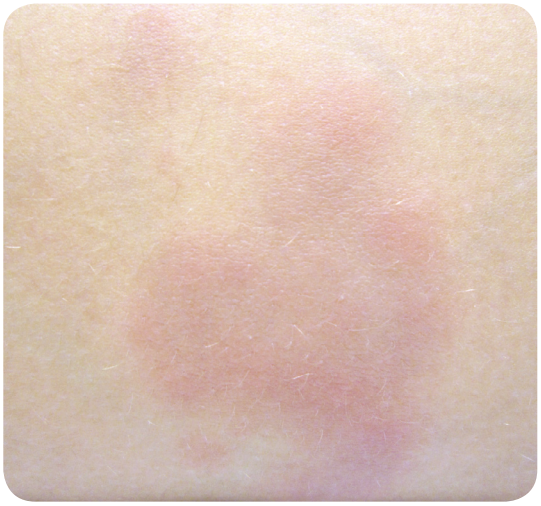

Figure 9.1 A single area of erythema nodosum.

DIAGNOSIS

- Observation of typical skin lesions

LAB RESULTS

- Biopsy in uncertain cases
- Additional evaluation to determine the underlying cause
  - Complete blood count, erythrocyte sedimentation rate, antistreptolysin-O titer, throat culture, urinalysis, intradermal tuberculin test, venereal disease research laboratory (VDRL), and cultures, as appropriate

DIAGNOSTIC IMAGING

Chest X-ray

- Additional evaluation to determine the underlying cause

TREATMENT

MEDICATIONS

- Potassium iodide, corticosteroids and colchicine can be used in severe refractory cases

OTHER INTERVENTIONS

- Address underlying cause
- Symptomatic management
  - Bedrest, leg elevation, compressive bandages, wet dressings, and nonsteroidal anti-inflammatory agents

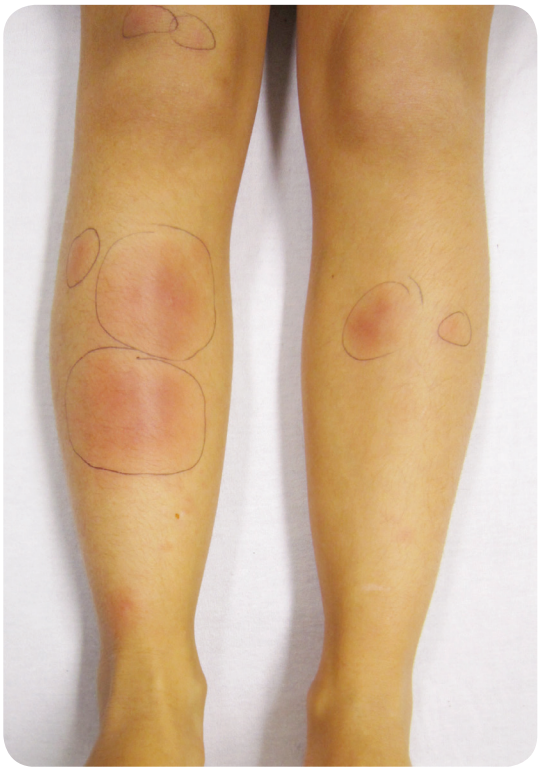

Figure 9.2 Erythema nodosum affecting the shins; a common site for this disease.

HEREDITARY ANGIOEDEMA (HAE)

osms.it/hereditary-angioedema

PATHOLOGY & CAUSES

- Small but important number of all cases of angioedema
  - Increased vasodilation and vascular permeability → fluid leakage from deep blood vessels → angioedema
  - Urticaria and pruritus are not present

CAUSES

- Inherited in an autosomal dominant manner involving mutation of genes associated with C1-inhibitor (*C1INH*) that inhibits the complement pathway and is associated with coagulation factor XII
  - Results in unregulated levels of bradykinin and other vasoactive substances → inflammation, vasodilation, and cellular injury
  - Attack triggers may include minor trauma, mood and temperature changes, but often no obvious inciting event can be established

SIGNS & SYMPTOMS

- Recurrent attacks of angioedema
- Painless, nonpruritic, nonpitting swelling of extremities, genitalia, buttocks, eyelids, lips, tongue, larynx or gastrointestinal tract
  - Gastrointestinal tract → nausea, vomiting, intense colicky abdominal pain, diarrhea, dehydration, and intense exhaustion → mimics a surgical emergency and unnecessary surgery could be performed
  - Larynx → life-threatening airway obstruction → without treatment, death by asphyxia occurs in about 25%
- Tightness, tingling, or erythema marginatum corresponding to the affected area may precede the swelling
- Each episode usually resolves within 72 hours
- Attacks begin during childhood and become increasingly frequent and severe
- Frequency of attacks differs greatly, varying from weekly episodes to intervals longer than a year; discrepancies can occur among different individuals and at different times in the same individual

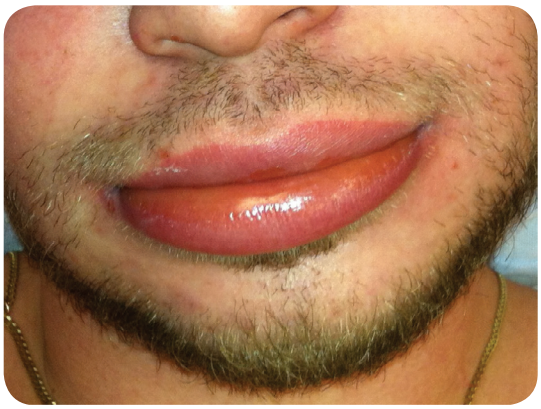

Figure 9.3 Angioedema of the lips.

DIAGNOSIS

DIAGNOSTIC IMAGING

- Imaging studies may be useful during attacks of gastrointestinal edema

LAB RESULTS

- Complement testing to assess alterations in the system

TREATMENT

MEDICATIONS

- Management of attacks
  - Intravenous C1-inhibitor concentrates, kallikrein inhibitors (ecallantide), bradykinin B2 receptor antagonists (icatibant) or, if those are unavailable, fresh-frozen plasma as an alternative

- More than one episode in a month or high risk of developing laryngeal edema → long-term prevention
  - Danazol (an androgen that increases levels of C4)
  - C1-inhibitor concentrates

OTHER INTERVENTIONS

- Avoid specific stimuli that have previously led to attacks
- Avoid medications associated with attacks
  - ACE inhibitors; medications containing estrogen

URTICARIA (HIVES)

osms.it/urticaria

PATHOLOGY & CAUSES

- Acute (< six weeks) or, rarely, chronic (> six weeks) skin eruption
- Acute form most common dermatologic disorder seen in emergency department
  - Most often benign and self-limiting, though may rarely progress to life-threatening angioedema or anaphylactic shock; strong tendency to recur
- Hypersensitivity reaction → mast cell degranulation and release of inflammatory mediators → increased vascular permeability → fluid leakage from superficial blood vessels → cutaneous lesion

TYPES

- Acute urticaria
  - Single lesions usually last less than 24 hours
- Chronic urticaria
  - May last six weeks or more

CAUSES

- Assessment for potential causes includes "5 Is"
  - Infection (bacterial/viral/fungal/parasitic)
  - Injection of a drug/insect venom
  - Inhaled substances (pollen, mold, dust)
  - Ingestion of foods, drugs, chemicals
  - Internal disease process such as an autoimmune disorder
- Vasculitis urticaria associated with autoimmune and malignant diseases
- Precipitants include psychological and physical stress, cold or hot temperature, pressure or vibration
- Physical urticaria is urticaria is induced by an exogenous physical stimulus such as scratching or firm stroking of the skin
  - The most common type of physical urticaria is called a dermatographism

SIGNS & SYMPTOMS

- *Wheals*: skin eruption characterized by itchy, burning or stinging, red, raised plaques with well-defined erythematous margins and pale centers
  - Individual lesions may coalesce
  - New lesions may appear as others resolve
- Can occur anywhere, but common sites are areas exposed to pressure (e.g., trunk, distal extremities, ears)

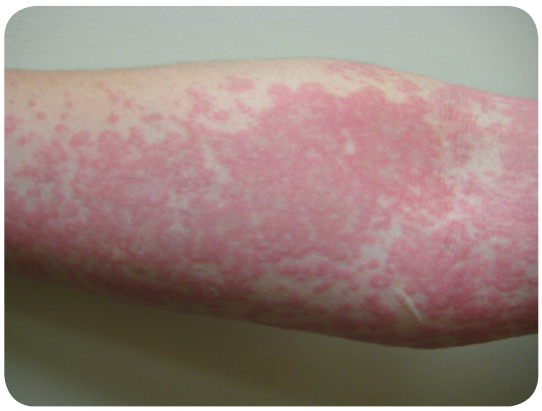

Figure 9.4 Urticaria of the forearm.

DIAGNOSIS

- Typically based on appearance
- Patch testing to confirm and determine the allergy

LAB RESULTS

- Complete blood count
- Erythrocyte sedimentation rate
- Thyroid-stimulating hormone (rule out thyroid disease)
- Autoimmune screening

TREATMENT

- Avoid triggers
- Symptomatic management
  - Antihistamines
  - In severe cases, corticosteroids or leukotriene inhibitors
  - Monoclonal antibodies and immunosuppressants may be used in refractory cases