

Urticaria - Angioedema Serum Sickness

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Urticaria -Angioedema : Etiology

- **Urticaria -Angioedema are best Considered as Symptoms**
- **Urticaria : Superficial Skin Layer (Dermis) Swelling**
- **Angioedema : Deep Skin Layer Edema (Nonpitting)**
- **Primary Effector Cell : Skin Residing Mast Cells**
- **Main Involved Mediators :**
 - **Histamine, Leukotrienes, Platelet-activating Factor**
 - **Prostaglandins , Cytokines**

Urticaria - Angioedema : Etiology

- **Common Triggers of Mast cell-Basophil Mediator Release ;**
 - **IgE Mediated : Food Allergens , Medications , Insect Venom**
 - **Anaphylatoxins (C3a-C5a) : Serum Sickness , Infections**
 - **Direct Pharmacologic Effect : Morphine , Radiocontrast**
 - **Physical-Mechanical : Dermatographism , Cholinergic Urticaria**

Urticaria - Angioedema : Classification

- **Acute Urticaria ;**
 - **IgE Mediated : Food Allergens , Medications**
 - **Viral Infections**
- **Chronic Urticaria (Persistence Of Symptoms Beyond 6 Weeks) ;**
 - **Physical Urticaria**
 - **Autoimmune Urticaria (IgG against IgE or IgE Receptor)**
 - **Chronic Spontaneous Urticaria (Formerly Idiopathic)**

Urticaria - Angioedema : Classification

- **Chronic Physical Urticaria ;**
 - **Dermatographism is the Most Common (Minimal Trauma)**
 - **Cholinergic Urticaria (Increase in Core Body Temperature)**
 - **Cold Urticaria-Angioedema**
 - **Solar Urticaria**
 - **Vibratory Urticaria**

Urticaria - Angioedema : Epidemiology

- **Acute Urticaria ; 15-25 % Of General Population**
- **Chronic Urticaria ; 1% Of Population**

Urticaria : Clinical Manifestations

- **Sudden Onset , Intensely Pruritic , Erythematous Skin Lesion**
- **Typically Resolve in 1-2 hr But Each Lesion May Last 24 hr**

Urticaria



Urticaria



Cholinergic Urticaria



IgE Mediated Drug Reaction

Angioedema : Clinical Manifestations

- **Deep Dermis - Subcutaneous Tissue Edema**
- **Essentially Nonpruritic**
- **Minimal Pressure-Pain Sensation**
- **Generally Last Longer than 24 hr**
- **Life-threatening Respiratory Obstruction is Possible**

Angioedema

Urticaria with Angioedema



Acute Urticaria - Angioedema : Lab. Evaluation

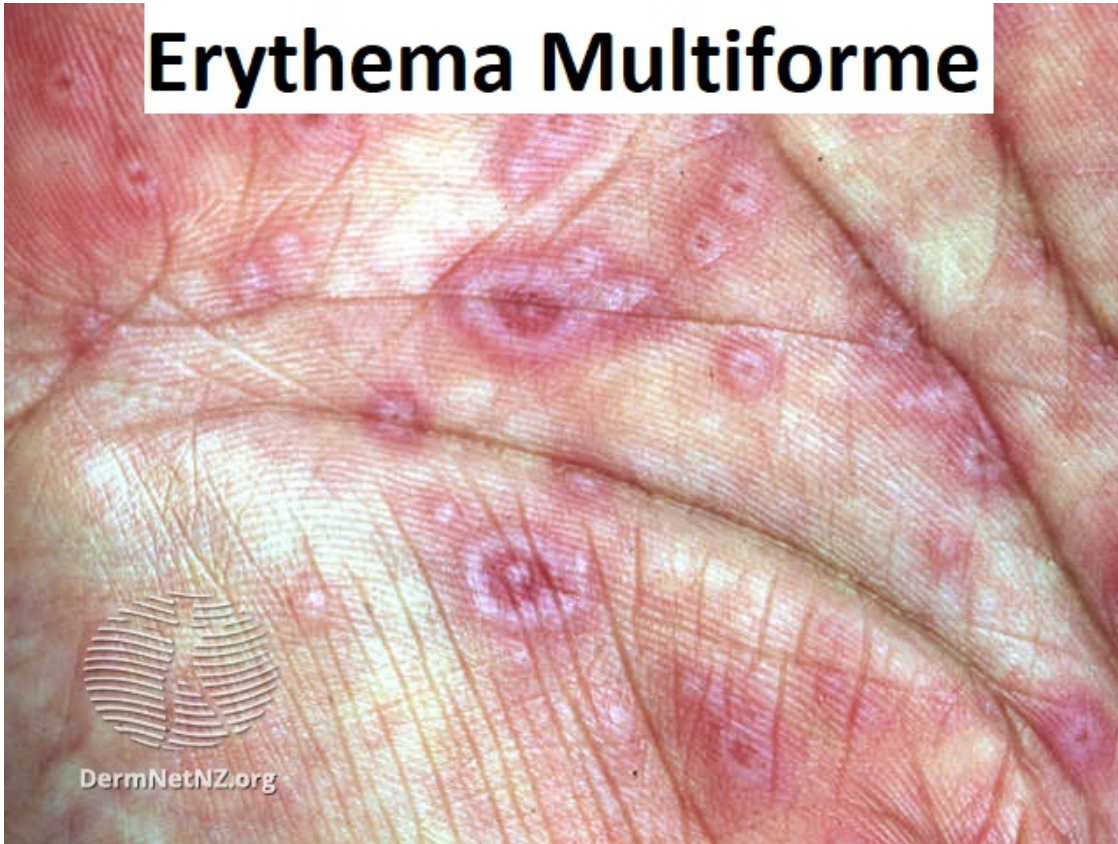
- **No Routine Studies Necessary**
- **Evaluate The Diagnosis You Suspect ;**
 - **IgE or Skin Test For Food Allergy**
 - **Skin Test For Hymenoptera Sting Allergy**
 - **Skin Test For Drug Allergy**
 - **Testing For Viral Infection if Necessary**

Urticaria - Angioedema : Differential Dx

- **Important Feature To Consider ;**
 - **Each Urticarial Lesion Lasts Under 24 hr**
 - **Urticarial Lesions are Pruritic (Not Painful)**
 - **Urticarial Lesion Blanch with Pressure**
 - **No Residual Skin Discoloration**

Urticaria - Angioedema : Differential Dx

Erythema Multiforme



Urticaria - Angioedema : Differential Dx

Dermatitis Herpetiformis



Urticaria - Angioedema : Differential Dx

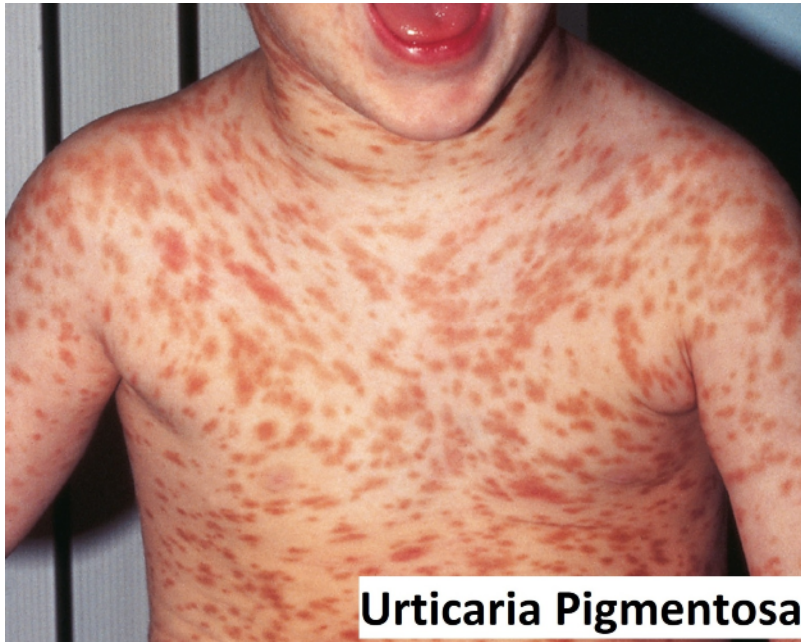


Bullous Pemphigoid

Urticaria - Angioedema : Differential Dx

- **Systemic Disease With Urticarial or Urticaria-Like Skin Lesion ;**
 - **Mastocytosis (Cutaneous , Systemic)**
 - **Muckle-Wells Syndrome**
 - **Schnitzler Syndrome**
 - **Urticarial Vasculitis**

Cutaneous Mastocytosis



Urticaria - Angioedema : Treatment

- **Acute Urticaria–Angioedema ;**
 - **Avoidance Of Triggering Agents**
 - **New Generation H1–Antihistamines**
 - **Allergist Referral For Documenting Food or Drug Allergy**
- **Chronic Urticaria ;**
 - **New Generation H1–Antihistamines (Up to 4 times Usual Dose)**
 - **Omalizumab , Cyclosporin if No Response To H1–Blocker**

Hereditary Angioedema

- **Unpredictable Recurrent Swelling Attack**
- **Extremities , Genitalia , Intra-Abdominal , Face , Pharynx**
 - **Some Patient Mistakenly Undergo Appendectomy**
- **No Urticaria - No Skin Lesion**
- **Trauma is a Common Trigger**
- **Bradykinin Pathway is The Main Culprit**
- **No Response To Antihistamines-Corticosteroid**

Hereditary Angioedema

- **Abnormal C1-Inhibitor (Level or Function) ;**
 - **Autosomal Dominant 75% , Sporadic 25%**
 - **Low C4 Level Even Between Attacks**
- **Normal C1-Inhibitor (Level & Function) ;**
 - **More Prevalent in Women**
 - **Mutation in Factor XII , Plasminogen , Angiotensin 1**

Angioedema



Hereditary A.E. : Diagnostic Evaluation

- **Screening Test : Complement C4 Level**
- **Evaluation of C1-Esterase inhibitor Level & Activity**
- **Genetic Study For Mutation Detection (SERPING1 , FXII , PLG , ANGP1)**

Hereditary Angioedema

- **Available Therapies ;**
 - **C1-Inhibitor Concentrate**
 - **Kallikrein Inhibitor**
 - **Bradykinin B2 Receptor Antagonist**
 - **Androgenic Steroids**
 - **Tranexamic Acid**

Serum Sickness

- **Type 3 Hypersensitivity Reaction (Immune-Complex Mediated)**
 - **Ag-Ab Complex Deposition and Tissue Injury (Vasculature , Kidney)**
- **Common Triggers ;**
 - **Blood Products**
 - **Foreign Proteins : Antithymocyte Globulin , Antivenoms, Antitoxins**
 - **Medications : Penicillin, Sulfonamides, Minocycline, Cephalosporins, Hydantoins, Thiazides**
 - **Viral Infections**

Serum Sickness : Clinical Manifestations

- **Symptom Begins 1–3 weeks After Allergen Exposure**
 - **1–4 Days if Patient Previously Exposed**
- **Classic Clinical Manifestations ;**
 - **Fever , Arthralgia–Arthritis–Myalgia , Skin Rash , Lymphadenopathy**
- **Other Rare Manifestations ;**
 - **Carditis, Glomerulonephritis, Guillain–Barre Syn., Encephalomyelitis, Peripheral Neuritis**

Serum Sickness : Clinical Manifestations



Serum Sickness : Clinical Manifestations



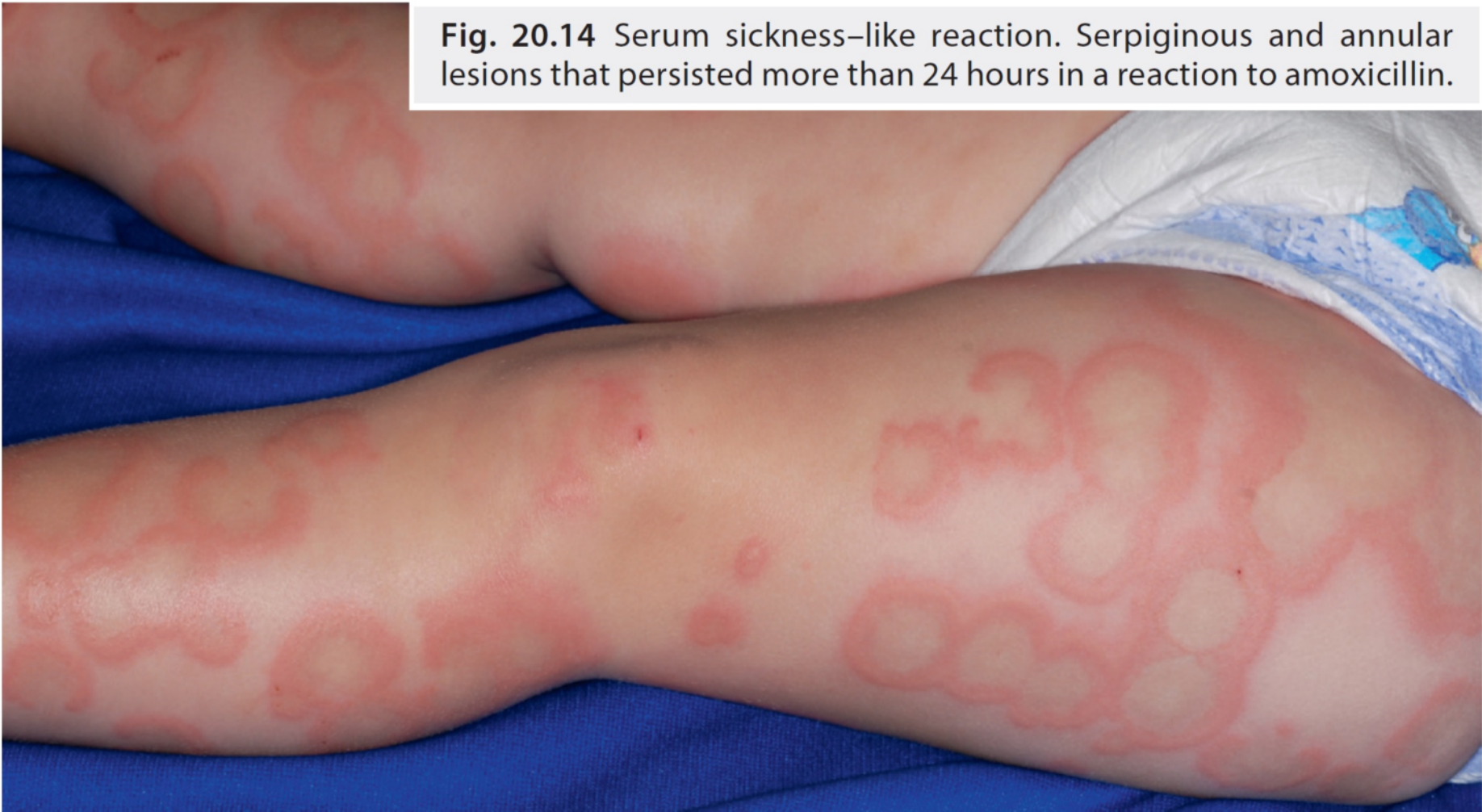
Fig. 20.13 Serum sickness–like reaction. Urticarial wheals occurred 2 weeks after exposure to cefaclor.



Fig. 20.15 Serum sickness–like reaction (SSLR). Note the central lilac discoloration, similar to that of erythema multiforme and typical of SSLR. This girl developed the reaction after 10 days of amoxicillin.

Serum Sickness : Clinical Manifestations

Fig. 20.14 Serum sickness–like reaction. Serpiginous and annular lesions that persisted more than 24 hours in a reaction to amoxicillin.



Serum Sickness : Clinical Manifestations



Fig. 20.16 Serum sickness–like reaction (SSLR). Note the swollen hand and large urticarial wheals in this girl with SSLR and arthralgias.

Serum Sickness : Differential Diagnosis

- **Viral Illnesses With Exanthems**
- **Kawasaki Disease**
- **Hypersensitivity Vasculitis**
- **Acute Rheumatic Fever**
- **Acute Meningococcal Or Gonococcal Infection**
- **Endocarditis**
- **Systemic-onset Juvenile Idiopathic Arthritis (Still Disease)**
- **Lyme Disease**
- **Hepatitis**
- **Other Types Of Drug Reactions**

Serum Sickness : Lab. Evaluation

- **Generally Done to Investigate Differential Diagnosis**
- **CBC diff ; Thrombocytopenia**
- **ESR - CRP ; Maybe Elevated**
- **U/A ; Mild Proteinuria , Microscopic Hematuria**
- **BUN-Cr , AST , ALT , Albumin**
- **CH50 , C3 , C4 ; Maybe Decreased - C3a ; Maybe Increased**
- **Testing For Viral - Bacterial Infections If Necessary**

Serum Sickness : Treatment

- **Treatment Is Primarily Supportive**
- **Antihistamines for Pruritus**
- **Acetaminophen For Fever**
- **NSAID for Mild Arthralgia-Myalgia**
- **Corticosteroids for Severe Disease ;**
 - **Fever over 38.5° c**
 - **Severe Arthralgia-Myalgia**
 - **Internal Organ Involvement (Nephritis , Carditis , ...)**

Final Exam Reference

- **Nelson Essential Of Pediatrics 8th Edition 2019**