Two Hour Visual Diagnosis Session
Patterned After

“What's Your Diagnosis”

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DISCLOSURE

I have no relevant financial relationships with commercial interests to disclose.

Case #1
7-Year-Old Female

- Admitted for severe rash.
- PMH – positive for similar rash waxing and waning since infancy, with some episodes of *Staph aureus* cellulitis.
- Exam revealed a generalized patchy erythematous rash with marked damage due to scratching.

Improved with intensification of topical therapy:
- Steroid cream
- Emollient
- Antihistamine

1-Yr. later: Now 8-Year-Old Female

- Readmitted for the same severe rash.
- Problem is thought to be severe atopic dermatitis related to poor compliance due to a very poor & difficult social situation; single, disabled parent.
- Again improved with intensification of topical therapy.
Now 11-Year-Old Female

◆ Three years later:
  – More skin inflammatory flares with several more episodes of cellulitis.
  – One episode of *Staph aureus* sepsis.
  – Septic hip and osteomyelitis.

Now 15-Year-Old Female

◆ Five years later:
  – Admitted for *Staph aureus* cellulitis of the right lower leg.
  – Follow up in the ID clinic revealed resolving cellulitis as well as several herpetic lesions:

15-Year-Old Female

◆ Several years earlier, the following immune screening studies showed:
  – Compliment = normal screen.
  – CBC normal except mild eosinophilia.
  – HIV = negative.
  – Oxidative burst assay = normal.
  – IgG response to vaccine antigens - NL
15-Year-Old Female

Several years earlier, the following immune screening studies showed:

- Quant. immunoglobulins (units / ml):
  - IgG = 1120 (614 – 1440)
  - IgM = 83 (48 – 333)
  - IgA = 214 (79 – 347)
  - IgE = 53,952 (0 – 260)
  - Repeat IgE 6-mo later = 103,682

What’s Your Diagnosis?

A. Bruton Syndrome
B. Job Syndrome
C. Common Variable Immune Deficiency
D. Severe Atopic Dermatitis

What’s Your Diagnosis?

B. Job Syndrome
- Davis, et.al., 1966, proposed the name due to tendency for Staph abscesses.
- Buckley, 1972, Hyper-IgE immune deficiency syndrome.
“Then Satan went out from the presence of the Lord and smote Job with boils from the sole of his feet to the crown of his head.”

The Book of Job, Chapter 2, verse 7
Most consistent with *Staph aureus* pyoderma & abscess.

Hyper-IgE Syndrome

CLINICAL FEATURES

- Atopic Dermatitis.
- Recurrent, often severe *Staph* infections of skin w/ 2º sepsis and bone and joint infections.
- Sinopulmonary infections.
- Mucocutaneous candidiasis.

Hyper-IgE Syndrome

- Pathogenesis is complex and incompletely understood (except by immunologists).
- Dysregulation of the immune system:
  - Th17 (T-helper 17 cell) defect appears key, along with other cytokines.
  - May have abnormal chemotaxis.
  - IgE levels thought to be part of the dysregulation rather than a direct cause for the increased frequency of infections.
Hyper-IgE Syndrome

Physical Features
- Coarse facial features.
- Frontal bossing.
- Broad nasal alar base.
- Tendency to resemble each other.
- Skin may reveal Lichenification.

Hyper-IgE Syndrome

Treatment:
- Meticulous care of skin – close the port-of-entry for bacteria (Staph).
- Early antimicrobial therapy for suspected infections; usually Staph.
- Involvement of an Immunologist; may try IVIG per NIH study.
- Genetic counseling.

Hyper-IgE Syndrome

Treatment:
- Meticulous care of skin – close the port-of-entry for bacteria:
  - Intensification of anti-inflamm. therapy and moisturizing.
  - Consider periodic decolonization.
  - Periodic prophylactic antibiotics; TMP / SMX, Doxycycline.
Hyper-IgE Syndrome

◆ Treatment:
  – Meticulous care of skin – close the port-of-entry for bacteria (Staph).
  – Early antimicrobial therapy for suspected infections; usually Staph.
  – Involvement of an Immunologist; may try IVIG per NIH study.
  – Genetic counseling.

Hyper-IgE Syndrome

◆ Treatment:
  – Genetic counseling:
    • Autosomal dominant:
      – Linked to mutations in the STAT3 gene (Signal Transducer and Activator of Transcription 3).
    • Autosomal recessive:
      – DoCk8 (Dedicator of Cytokinesis 8).

Hyper-IgE Syndrome

◆ Great Reference:
  – Timothy R. LaPine & Harry R. Hill of the University of Utah School of Medicine, Up To Date.
What's Your Diagnosis?

A. Bruton Syndrome  
   - COL Ogden Bruton @ Walter Reed, described in 1952 in an 8-year-old male.  
   - X-Linked (♂ not in females) mutation of Bruton Tyrosine Kinase (BTK) gene, results in blocking B cell development.  
   - Treat with IVIG replacement every 3 – 4 weeks.  
   - Early treatment of infections.  
   - Pneumococcal immunizations.

What's Your Diagnosis?

C. Common Variable Immune Deficiency  
   - ≈ 10% of all primary deficiencies.  
   - Low IgM, IgA & IgG (not as severe as Bruton).  
   - Variable severity – recurrent infections, autoimmune disease, lymphoid malignancy.  
   - Treat with IVIG – individualized.
What’s Your Diagnosis?

D. Severe Atopic Dermatitis
- May also have increased frequency of infections from barrier defect (broken).
- Screen patients for Hyper-IgE if not responsive to common therapies or if recurrent infections are occurring.
- Also think of Langerhans-cell Histiocytosis

What’s Your Diagnosis?

D. Severe Atopic Dermatitis
- Eczema herpeticum can occur with any chronic skin disorder; not necessarily due to hyperimmunoglobulin E syndrome.

9 Year-Old Female
- Fell while running, impaling a roofing nail into her left knee.
- Immunizations: UTD by History
9 Year-Old Female

- In the ED, radiographs were taken.

Nail removed and wound cleaned; sent home on Cephalexin; 3-days later:

- Admitted for IV anti-Staph antibiotics.
- At that time, written documentation of immunizations were provided:
  - 4 doses of tetanus toxoid between 2 and 18 months of age.
What Tetanus Prophylaxis Would You Give?

A. None
B. Tetanus Immune Globulin (TIG)
C. Tetanus Toxoid
D. Tetanus Toxoid plus TIG

Tetanus Guidelines

◆ The only time TIG is recommended is when < 3 doses of toxoid have been given, AND it is a tetanus-prone injury:
  - Contaminated with dirt / feces / saliva
  - Puncture wounds
  - Avulsions, crush, burn, frostbite injuries
◆ Toxoid is recommended if < 3 doses, or if > 5 years since last dose, AND if tetanus-prone or > 10 years if not.

9 Year-Old Female

◆ Had 4 previous doses, but > 5 years since last dose, AND tetanus-prone.
◆ She was treated with a dose of Tdap (≥ 7 years old), and IV Clindamycin for a couple of days with good results and sent home on PO Clindamycin.
Case #3

Another Quickie.

5-Day-Old Female

- To the ER with a swollen left eye.
- Onset was earlier that morning.
- Eyelids were stuck closed with dried exudate.
- VS – normal.
5-Day-Old Female
- Mother’s P, L, Delivery – normal with good prenatal care.
- No history of sexually-transmitted infections w/ neg prenatal screening.
- Jaundice-only postnatal complication.
- Exam – normal baby, except for scleral icterus, conjunctival and preseptal skin erythema of right eye.

5-Day-Old Female
- PCR testing was negative for:
  - HSV, Gonorrhea, Chlamydia, and Adenovirus (not FDA-approved).
- CBC – normal.
- Blood culture – negative at 48 hours.
- Gram stain – mixed organisms.
- Culture – H. influenzae (non-typable).

What’s Your Diagnosis?
A. Haemophilus influenzae conjunctivitis
B. Chlamydia conjunctivitis
C. Escherichia coli conjunctivitis
D. Vitamin A deficiency
**What’s Your Diagnosis?**

A. *Haemophilus influenzae* conjunctivitis
   - Considering all-comers, it’s 2nd only to *C. trachomatis* as bacterial causes at this age.
   - Usually responsive to topical drops or ointment containing an aminoglycoside or polymyxin B for about 5 days.

**What’s Your Diagnosis?**

B. Chlamydia conjunctivitis
   - *Chlamydia trachomatis* very unlikely in this case:
     • Good prenatal care.
     • No other STI’s in mother.
     • Negative PCR (even though not official).
   - May be accompanied by pneumonia.

**Chlamydia Conjunctivitis**

◆ *C. trachomatis* – most common STI.
◆ Infant colonized during delivery.
◆ Infection between 3 days – 3 weeks.
◆ Dx often clinical, supported by positive maternal test. PCR of eye discharge not yet FDA-approved.
◆ Tx – Erythromycin X 14 days - ?PS
◆ Azithromycin X 3 days may be option.
Trachoma

◆ Chronic / recurrent infection with scarring and neovascularization.

What’s Your Diagnosis?

◆ Escherichia coli conjunctivitis
  – Not a likely cause of conjunctivitis.
  ❖ However, it appears to be displacing group B strep as the most common cause of neonatal sepsis and meningitis.

  ❖ Greenhow, et. al., *Pediatrics* 2012;129;e590.

What’s Your Diagnosis?

D. Vitamin A deficiency – if severe:
  – Retinopathy – night blindness.
  – Severe dryness / conjunctival thickening (xerophthalmia).
  – Eventual Blindness.
  – This degree of malnutrition is fairly rare in this country.
Vitamin A Deficiency Effects on the Immune System

◆ Most impact is on the cellular system, but interferes with both the Cellular and Humoral systems of mucosal immunity:
  – More respiratory and GI infections.
◆ Severe measles.

Bilateral Conjunctivitis

◆ If seen on the first day-of-life, think chemical conjunctivitis (prophylaxis).
◆ Should raise suspicion for a viral etiology; Adenovirus most common.
◆ More likely to be contagious among family members and close contacts.
◆ Don’t forget to ask or look at parents and siblings.
Case #4
Complicated, yet Simple.

14-Year-Old Male with Orbital Mass
◆ HPI - Onset 1-week earlier with nausea, vomiting and headache:
◆ After 3 days he was seen in small hospital ER & Dx with a migraine.
◆ A couple of days later, his symptoms persisted & he developed dysconjugate gaze and he went to a 2nd hospital where a CT was done, showing something in the right orbit.

14-Year-Old Male with Orbital Mass
◆ HPI - Onset 1-week earlier with nausea, vomiting and headache:
   – Full sepsis work up – normal.
   – Vanc + Ceftriax started – transferred.
14-Year-Old Male with Orbital Mass

◆ Past medical history:
  – Previously healthy – imm. UTD
  – Staph infection of knee several weeks earlier with several superficial abscesses drained by primary and Tx empirically with TMP / SMX (no cult).
    • Knee still has evidence of old infection.

◆ Family history:
  – Mother was also treated for a skin abscess around the same time.

◆ Examination:
  – Normal vital signs (no fever).
  – Knee findings previously noted.
  – Right eye a bit injected with dysconjugate gaze.
14-Year-Old Male with Orbital Mass

Lab tests:
- CBC = normal

MRI:
- Right orbital mass.
14-Year-Old Male with Orbital Mass

- Due to no fever over 3 days and normal CBC, and an unclear MRI reading, Ophthalmology consultant diagnosed: “Orbital Pseudotumor”
- Discharged on oral steroids and Clindamycin (just in case) with initial improvement in pain.

9 Days Later, readmitted with return of pain, and getting worse.
- CBC now = 20,000 WBC’s w/ 67% Gran.
- Repeat MRI:
  - This time it was read as an abscess.
  - Had surgical drainage on the same day revealing pus.
What’s Your Diagnosis?
A. Group A Streptococcus
B. Staphylococcus aureus
C. Haemophilus influenzae b
D. Streptococcus pneumoniae

What’s Your Diagnosis?
B. Staphylococcus aureus (MRSA)
  – Most common overall.
  – Recent multiple skin abscesses.
  – Mother had recent skin infection.
  – Most likely caused by hematogenous seeding from the knee infection a few weeks earlier.

What’s Your Diagnosis?
B. Staphylococcus aureus (MRSA)
  – One Additional Teaching Point:
    Should not be fooled by progression in spite of
    “just in case” Clindamycin which, in a case like this, is
    mere folly!
What’s Your Diagnosis?
◆ Orbital Pseudotumor
   – AKA idiopathic orbital inflammatory syndrome.
   – Can present as an orbital mass.
   – Main treatment is with steroids.

14-Year-Old Male with Orbital Mass
◆ The patient did well with one month of IV Clindamycin (700 mg q8h), and one month high-dose oral Clindamycin at 600mg TID.

Case #5
Another Unusual Case I Had Never Seen Before.
13-Year-Old Male

- Admitted with pneumonia, fever, sore throat and respiratory distress.
- Onset 3-days earlier with flu-like Sx.
- Was taken to urgent care clinic where he was clinically diagnosed with strep throat, based on erythema and petechiae in the throat (no culture), and pneumonia by CXR:

He was given Bicillin (for Strep?) and transferred for admission.

He was empirically continued on antibiotics (Ceftriaxone).

PMHx:
- Healthy except for mild asthma.
- Immunizations documented UTD.
- No preceding illness or medicines.
13-Year-Old Male

◆ Family Hx:
  – Mother recently had a URI.
  – Father recently returned from Afghanistan, but is well.
◆ Examination: T = 101.9°F, RR = 30
  – Soon after admission, mouth lesions worsened with general mucousitis.
  – Eyes, lips and urethra also red.

13-Year-Old Male

◆ Lab tests:
  – Admission RVP + for Rhinovirus.
  – Other tests, including Mycoplasma IgG and IgM titers were negative.
◆ HINT: 7-days later, repeat *Mycoplasma pneumoniae* IgG mildly elevated and IgM strongly elevated.
What's Your Diagnosis?
A. Atypical Stevens-Johnson Syndrome
   - Severe oral mucousitis
   - Conjunctivitis
   - Cracked, bleeding lips
   - Urethritis / vaginitis
   - Minimal to no skin manifestations

What's Your Diagnosis?
B. Kawasaki Syndrome
   - 13 years is a bit old for this diagnosis.
   - The limited rash and mucous membrane findings do not fit; more consistent with erythema multiforme.
Kawasaki Syndrome

5 Days of Fever, plus
Must have 4 of the 5 diagnostic criteria:
1. Conjunctivitis without discharge.
2. Polymorphous Rash.
3. Extremity changes.
4. Other mucous membrane involvement – inflammation of mouth, urethra, vagina, anus.
5. Lymphadenopathy (>1.5 cm diameter)

Erythema Multiforme

◆ Mostly round or oval, fixed erythematous lesions.
◆ Progresses to form concentric zones of epidermal injury with color change to form “target” lesions; 1 – 3 weeks.
◆ Palms and soles frequently involved.
◆ Often triggered by Herpes simplex.
◆ Frequently confused with Urticaria.
Erythema Multiforme

- Major = at least 2 mucous membrane surfaces (like S - J Syndrome).
- Minor = one or no mucous membrane surface involvement.
- The above classification is not widely used anymore, but rather:
  - E. M. minor = E. multiforme.
  - E. M. major = S - J Syndrome.

Toxic Epidermal Necrolysis?

- Usually Preceding medications (anticonvulsants, sulfas, etc.).
- Severe, blistering rash.
- Mucous membrane involvement.
- Deeper cutaneous injury:
  - Subepidermal
- Treat like a burn.
Case #6

9½-Year-Old Male

- Bitten in the face by a fully vaccinated Rottweiler in a provoked attack.
- To Community ER – 2 puncture wounds; under the right eye and over right maxilla.
- The patient’s immunizations were thought to be UTD; the father thought the last tetanus was 4 or 5 years ago; not sure.
- Wounds were cleaned and Rx Augmentin; but did not fill it.

The NEXT DAY, he was back in the ER with fever to 103°F.
- The skin around the wounds appeared swollen and erythematous.
- Sutures removed; given IV Clindamycin and told to take the Augmentin, but only took 1 dose before he was back with rapidly worsening pain and swelling.
- In the ER – IM Ceftriaxone & admitted.
9½-Year-Old Male

- Vital Signs – normal; Exam as shown:
  - Rest Normal.
  - Culture Pending.

What's Your Diagnosis?

2- Parts

Part 1

Does He Need a Tetanus Booster?

A. YES
B. NO
Does He Need a Tetanus Booster?
A. YES
   - There’s no documentation available.
   - Father’s not sure if it has been 4 or 5 years since his last dose.
   - Puncture (bite) wounds should be considered Tetanus-prone.
   - See case #2 discussion.

What’s Your Diagnosis?
Part 2

What’s Your Diagnosis?
A. *Staphylococcus aureus*
B. *Eikenella corrodens*
C. *Group A Streptococcus*
D. *Pasteurella multocida*
What’s Your Diagnosis?

C. *Group A Streptococcus*
- Culture positive.
- Predicted by rapid onset and speed of progression.
- Some overlap, and exceptions can occur, so you still need a culture.

What’s Your Diagnosis?

A. *Staphylococcus aureus*
- Most common overall.
- Usually > 48 hours in developing.
- Usually not as severe or invasive; more likely to get “walled off”.

What’s Your Diagnosis?

B. *Eikenella corrodens*
- Gram-negative, facultative anaerobe.
- Common cause of human bite infections, including clinched fist.
- Less common in animal mouths.
- Slow in developing; may take up to a week or more to manifest.
What’s Your Diagnosis?

D. Pasteurella multocida
- Gram-negative coccobacillus.
- Commonly found in cat bites.
- Similar features as group A strep.
- DOC = any penicillin.

Case #7
32-Week Preterm Newborn Female

- Prolonged ROM with C-S delivery due to failure to progress.
- Maternal fever with chorioamnionitis.
- Pregnancy and PMHx – normal.
- Prenatal screening all negative.
- No Hx of STI’s (STD’s), including herpes.
- Examination of baby was normal.
- BC sent and Amp + Gent started.

32-Week Preterm Newborn Female

- Baby did well till 5-days of age:
  - Noted some blistering sores on her low back as shown - - -

What’s Your Diagnosis?

A. Neonatal Lupus
B. Neonatal Herpes
C. Langerhans Cell Histiocytosis
D. Neonatal Candidiasis
What’s Your Diagnosis?

B. Neonatal Herpes
- Not a “brain-buster”.
- The appearance is right.
- The timing is right:
  - Usually appears \( \approx 5 - 14 \) days.
  - Diagnosed with PCR of the lesion.

What’s Your Diagnosis?

B. Neonatal Herpes
- Baby also had a full sepsis work up:
  - Blood culture and HSV PCR
  - CSF analysis with HSV PCR
  - Liver enzymes
  - Chest radiograph
- All negative \( \therefore \) Skin Eye Mouth (SEM)

What’s Your Diagnosis?

B. Neonatal Herpes
- Baby treated with IV Acyclovir at 60 mg/kg/day \( \div q 8 \) hours, for 14 days, followed by PO acyclovir at 300 mg/M\(^2\) every 8 hours for prophylaxis for 6 months; monitoring for neutropenia.
- Improved neurodevelopmental outcome.

Neonatal Herpes Exposure?

- New Guidelines for managing the newborn of a mother with known or suspected HSV around time of delivery.
- Hinges significantly on maternal history of HSV.
- Also new is the Herpes PCR in blood.

Entire guideline statement can be found at http://pediatrics.aappublications.org/content/131/2/e635.full.pdf

Print out the two-page algorithm for ready reference in the nursery work area.

Neonatal Lupus

- Mothers with Lupus or Sjögren syndrome with circulating autoantibodies.
- Typical discoid, annular, erythematous plaques with central scale.
- May have heart block that may persist.
Neonatal Lupus

- Skin lesions resolve when mother’s autoantibodies clear.
- No treatment needed unless a pacemaker is required.

What’s Your Diagnosis?

C. Langerhans Cell Histiocytosis
- May be seen in newborns.
- Various manifestations:
  - Discrete plaque-like lesions.
  - Papulo-nodular lesions.
  - Diffuse, persistent diaper rash-like appearance.

Langerhans Cell Histiocytosis

- Paul Langerhans (1847 – 1888) – German Physician – Pathologist.
- Formerly Histiocytosis X.
- Three subtypes (much overlap):
  1. Letterer-Siwe disease
  2. Hand-Schüller-Christian disease
  3. Eosinophilic Granuloma

Of Historic Interest Only.
Langerhans Cell Histiocytosis

- Letterer-Siwe disease
  - Most severe; often fatal.
  - Multi-organ involvement.
  - Persistent “diaper rash”.
  - Bone lesions.
  - Tx with chemotherapy.
  - Death by respiratory failure.

Langerhans Cell Histiocytosis

- Hand-Schüller-Christian disease
  - Exophthalmos
  - Diabetes insipidus
  - Lytic skull lesions

Langerhans Cell Histiocytosis

- Eosinophilic Granuloma
  - Lytic bone lesions
  - Usually skull
What’s Your Diagnosis?

D. Neonatal Candidiasis
   – Appearance a bit different than HSV.
   – Timing usually earlier than HSV.
   – Fungal stain of pustule should be + for pseudohyphae.
   – TX depends on severity:
     • Amphotericin b or Fluconazole.

Case #8

Something a Bit More Exotic?

Thanks to Roberto P. Santos, M.D.
Pediatric Infectious Diseases
Albany Medical Center
Albany, New York
4-Year-Old Male
◆ To primary for evaluation of lesions on his cheek, hand and flank area.
◆ First noticed on face and hand while visiting relatives in Iran several weeks earlier.
◆ Treated locally with cryotherapy without much improvement – returned to U.S.
◆ PMHx – previously healthy; no prior skin problems or sick contacts.

Rest of exam is normal.

What’s Your Diagnosis?
A. New World Cutaneous Leishmaniasis
B. Cutaneous Myiasis
C. Old World Leishmaniasis
D. Leprosy
What's Your Diagnosis?

C. Old World Leishmaniasis
- Biopsy + for *Leishmania major*.
- Obligate intracellular parasite that targets the mononuclear phagocytes
- Transmitted by the bite of the female sand fly:

LEISHMANIASIS
THREE CLINICAL SYNDROMES

1. Cutaneous - inoculation – enter into mononuclear phagocytes - erythematous nodule - ulcer w/ raised borders.
3. Visceral (Kala-azar)
LEISHMANIASIS
THREE CLINICAL SYNDROMES

1. Cutaneous - inoculation - into mononuclear phagocytes - erythematous nodule - ulcer w/ raised borders.
3. Visceral (Kala-azar) - tropical splenomegaly

Leishmaniasis Treatment
◆ Individualize; may not need treatment.
◆ A Pentavalent Antimonial (Stibogluconate)
◆ Liposomal Amphotericin B
◆ Paromomycin
◆ Azoles
Leishmaniasis Treatment
◆ In March, 2014, CDC approved Miltefosine, a phosphocholine analogue, for visceral leishmaniasis.
◆ Recommendations change; Consult CDC (770-488-7775).

What's Your Diagnosis?
A. New World Cutaneous Leishmaniasis
   – Similar diseases; different species:
     • L. mexicana, L. amazonensis, L. braziliensis, and others.
   – Similar treatment.

What's Your Diagnosis?
A. New World Cutaneous Leishmaniasis
B. Cutaneous Myiasis
C. Old World Leishmaniasis
D. Leprosy
What's Your Diagnosis?

B. Cutaneous Myiasis
   - “Skin maggots”

Relatively Harmless.

What’s Your Diagnosis?

A. New World Cutaneous Leishmaniasis
B. Cutaneous Myiasis
C. Old World Leishmaniasis
D. Leprosy
What’s Your Diagnosis?

D. Leprosy

- *Mycobacterium leprae*
- Infects skin, mucous membranes, testes and due to tropism for peripheral nerves, the lesions are painless, maybe numb and may be injured without knowing it.
- 5 histologic classifications:
  - Tuberculoid to Lepromatous.

What’s Your Diagnosis?

D. Leprosy

- Often acquired from family member.
- Dx – biopsy
  - PCR may be available.
- Tx: 24 mo. of Dapsone + Rifampin +/- Clofazimine if severe.
  
  Consult NHDP (800-642-2477)

Case #9

Not So Exotic.
11-Year-Old Female

- Admitted with painful swelling and erythema of her right foot.
- HPI:
  - 3-days earlier, she was running through her house, stepping on an overturned glass aquarium, that was not being used; breaking through the glass & jerking her foot up, sticking a shard of glass into it.

11-Year-Old Female

- HPI (continued):
  - The aquarium had been used for fish, then later to house a guinea pig. It had not been cleaned out.
  - Pain and swelling the next day & taken to urgent care – wound was cleaned and given Tdap + Rx for TMP / SMX (Septra®).
  - Returned the next day getting worse.

11-Year-Old Female

- Past medical history:
  - Healthy female.
  - Immunizations UTD.
  - The day prior to the accident, she had just finished a 10 day course of Amoxicillin for group A strep tonsillitis.
11-Year-Old Female

◆ Examination:
  – Normal vital signs and exam, except for the chief complaint:
    • Diffuse swelling of lateral foot.
    • Puncture wound as shown.
    • Pain and erythema just proximal to the 5th toe.

What's Your Diagnosis?

A. Staphylococcus aureus
B. Pseudomonas aeruginosa
C. Group A Strep
D. Mycobacterium marinum

In My Opinion
What’s Your Diagnosis?

C. Group A Strep
- Not culture proven – no drainage.
- Developed very quickly (< 24 hours).
- Connection to recent tonsillitis?
- Your guess is as good as mine.

What’s Your Diagnosis?

A. Staphylococcus aureus
- Could certainly be Staph aureus.
- Just a matter of timing.
- Usually takes a bit longer to present.
- Usually marked by abscess formation with yellowish pus.

What’s Your Diagnosis?

B. Pseudomonas aeruginosa
- Usually seen with puncture wounds through sneakers, 5 – 7 days later.
- Osteochondritis.
- Treat with surgical debridement and brief course of IV anti-pseudomonas antimicrobial; 7 – 10 days.
- May be a role for a Quinolone.
What's Your Diagnosis?

D. *Mycobacterium marinum*
- Nontuberculous mycobacterium.
- Causes cutaneous lesions after contamination of an injury in water containing the organism.
- Tx – none to surgical debridement, with or without medications:
  - Doxy, TMP/SMX, Clarithromycin.

11-Year-Old Female

◆ Treated with Clindamycin for a total of 7 weeks IV and PO with a good outcome.
◆ The osteomyelitis at such a distance from the puncture wound is explained by the mechanics of the injury.

As she jerks her foot back, the shard of glass takes this path; inoculating the bone.
And Finally
Case #10

14-Year-Old Female
◆ Referred by her dermatologist for eval of a + PPD.
◆ Was being treated for chronic, severe ulceronecrotic PLEVA; due to failing conventional Tx with PUVA and Doxy, she was about to begin Methotrexate.
◆ PMHx – immigrated from Pakistan about 7 months earlier; had a negative PPD at that time. The rash began a few weeks after arrival to the U.S.

14-Year-Old Female
◆ She had a healthy childhood except for being diagnosed with some sort of rheumatic disease a couple of years earlier (no records), but was on no Tx.
◆ She is also currently being followed by a rheumatologist for some joint pains, but no diagnosis.
◆ Examination showed many hyperpigmented ulcerative lesions - -
Pityriasis Lichenoides Et Varioliformis Acuta.

14-Year-Old Female
◆ The rest of her exam was normal.
◆ She had no signs or symptoms of TB; no fevers, cough, night sweats, weight loss, clear CXR; and no joint swelling.
◆ She had INH therapy started.
◆ Lab tests: numerous special rheum markers, inflammatory markers, metabolic tests, CBC’s, were all negative or normal.

14-Year-Old Female
◆ A lesion biopsy revealed granulomatous inflammation with epithelioid histiocytes, dermal necrosis, including some hair follicle necrosis.
◆ AFB stains and cultures, as well as M. tuberculosis PCR of the biopsy material, were negative.
14-Year-Old Female

◆ The rash soon began to improve, and by the end of her 9-months of INH, it had almost resolved according to her dermatologist, but no pictures.

    However,

◆ Within 6 months, she was back in the ID clinic with a recurrence of the rash:

What’s Your Diagnosis?

A. Langerhans Cell Histiocytosis
B. Severe PLEVA
C. Papulonecrotic tuberculid
D. Henoch-Schönlein Purpura
What’s Your Diagnosis?
C. Papulonecrotic tuberculid
- I know; I've never seen this either.
- It stumped her Dermatologist and Rheumatologist as well, until the Biopsy interpretation:

Pathologist Report
Dr. Brien,
In this clinical setting, the following constellation of findings make this biopsy compatible with papulonecrotic tuberculid:
1. Erosion /superficial ulceration of the epidermis.
2. Area of dermal necrosis in a "V" shape distribution.
3. Surrounding granulomatous inflammation.
Also, one study found hair follicle necrosis in 20% of papulonecrotic tuberculid; a finding that was also present in this case.
Hope this helps.

What’s Your Diagnosis?
C. Papulonecrotic tuberculid
- Hypersensitivity to TB infection, with or without active disease.
- Results in vasculitis by immune complex deposition or by subacute lymphohistiocytic vasculitis.
- As per Biopsy, necrosis is common result with granulomatous lesions containing epithelioid histiocytes.
What’s Your Diagnosis?

C. Papulonecrotic tuberculid
- Typically, no TB can be found in the granuloma.

- Treatment recommendation is same as for pulmonary TB for 6 months:
  - INH + Rifampin X 6 months
  - Ethambutol X 2 months
  - Pyrazinamide X 2 months

Recommend consulting a TB expert, and co-manage with a Dermatologist.
What's Your Diagnosis?
C. Papulonecrotic tuberculid
   - Lastly, with the chronic rheumatoid arthritis-type joint symptoms makes one wonder about Poncet Disease:
     • Reactive arthritis due to TB.
     • Normally see in active TB, not LTBI.
     • Since treatment, joints also improved; makes one wonder.

What's Your Diagnosis?
B. Severe PLEVA
   - Pityriasis Lichenoides Et Varioliformis Acuta.
   - Characterized by erythematous, scaly papules, rarely accompanied by hemorrhagic and papulonecrotic lesions.

PLEVA
◆ Victor Muca - 1877-1919
◆ Rudolph Habermann - 1884-1941
◆ Frequently Dx as “Atypical Varicella”
◆ Not an infectious disease
PLEVA

◆ Papulosquamous disorder.
◆ AKA Acute Lichenoid Pityriasis.
◆ Progresses through stages much like varicella.

CONTINUES FOR UP TO 9 MONTHS.
◆ Patient is not sick.
◆ Usually treat with just reassurance.
◆ If severe, a dermatologist may use PUVA therapy & or Doxycycline, and ultimately, Methotrexate.
◆ Most case are fairly mild.
### What's Your Diagnosis?

**D. Henoch-Schönlein Purpura**

![Image of skin rash]

### Henoch-Schönlein Purpura

- First recognized as a distinct syndrome by William Heberden (1710-1811) in the 18th century.
- Johann Schönlein (1793-1864) described the rash in 1837.
- Edouard Henoch (1829-1910) described the GI & Renal manifestations in 1874.

<table>
<thead>
<tr>
<th>Henoch-Schönlein Purpura</th>
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<tr>
<td>IgA-mediated autoimmune small vessel vasculitis.</td>
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<td>Most common between 2 – 8 years.</td>
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<td>Males about 2 X more than Females.</td>
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<td>Triggered by numerous infections.</td>
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Henoch-Schönlein Purpura

- Diagnosis by recognition of characteristic rash with one or more associated findings.
  - Rash -------------- % 100
  - Arthritis ------------ % 75
  - GI pain ------------ % 50
  - Renal involvement - - % 50*
  - CNS involvement - - Rare*

* These can be lethal.

Treatment is usually supportive if confined to the skin; occasionally with steroids or other non-steroidal anti-inflammatory drugs for GI symptoms, renal, or CNS disease; but no convincing evidence of benefit.

THE END