Avoiding Rash Decisions in the Work Up of a Patient with Refusal to Walk

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Disclosures
I have no disclosures
Presentation

Hospital Emergency Department

• 8 year old African American male with 3 months of left knee pain
• No trauma, no swelling, no fevers
• Previously treated with ibuprofen
• Now refusal to bear weight and new fevers and a rash on his abdomen and shins

History

• Past medical History
  • Hemoglobin C trait
  • Recent Strep infection 4 weeks ago – treated with azithromycin (allergic to penicillin)
  • Seasonal allergies
• Medications
  • Loratadine
  • Fluticasone intranasal
• Allergies
  • Amoxicillin - hives
  • Ofloxacin - hives
  • Penicillin - hives

• Family History
  • Sickle cell, Sarcoidosis, Hypertension - maternal grandmother
  • Rheumatoid arthritis and Thyroid disease - maternal aunt
  • Diabetes mellitus 2 - paternal grandfather
Physical Exam

Vitals: BP 103/59, T: 101.7°F (38.7°C), RR: 22, SpO2: 97% (RA)

- Height 141 cm (97%), Weight 36 kg (95%), Body mass index is 18.11 kg/(m^2) (85 percentile).
- General: anxious
- Heart: normal rate, regular rhythm, no murmur
- Lungs: CTA bilaterally no wheezes rhonchi or crackles
- Neurological: alert and oriented, moving all extremities but not against resistance, normal reflexes, unable to assess gait as he refuses to walk

Musculoskeletal and Skin Exam

- Left knee is painful when hip is flexed against resistance and knee extension against resistance. Patient refuses to stand and walk.
- Full range of motion, and no swelling or erythema of any joints. No weakness in either leg knee or foot.
# Differential

<table>
<thead>
<tr>
<th>Infectious</th>
<th>Rheumatologic</th>
<th>Heme/Onc</th>
</tr>
</thead>
<tbody>
<tr>
<td>Septic joint</td>
<td>Reactive Arthritis</td>
<td>Lymphoma</td>
</tr>
<tr>
<td>Rheumatic Fever</td>
<td>Systemic JIA</td>
<td>Histiocytosis</td>
</tr>
<tr>
<td>Lyme disease</td>
<td>Rheumatic fever</td>
<td>Orthopedic</td>
</tr>
<tr>
<td>Osteomyelitis</td>
<td>Lupus Vasculitis</td>
<td>Fracture</td>
</tr>
<tr>
<td></td>
<td>Kawasaki</td>
<td>Dislocation</td>
</tr>
<tr>
<td></td>
<td>Granulomatous disease</td>
<td>Trauma</td>
</tr>
<tr>
<td></td>
<td>Sarcoidosis</td>
<td>Growing pains</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Legg-calve-perthes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Avascular necrosis</td>
</tr>
</tbody>
</table>

## Which category on our differential is most concerning initially?

1. Infectious
2. Hematologic
3. Rheumatologic
4. Orthopedic

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Initial Work Up

Focus was on potential infectious causes

- ESR: 40
- CRP: 7.5
- CK: 96

- MCV: 71.5
- Neutrophil: 72.3%
- Lymph: 10.4%
- Mono: 13.7%
- Eos: 3%

UA: no nitrate, leukocyte esterase or blood or protein
Hip X-ray

Femoral heads are round and smooth without fragmentation or sclerosis. Epiphysis are appropriately aligned. No subluxation or dislocation. No fracture.

MRI of Leg

No evidence of osteomyelitis, subperiosteal abscess or drainable fluid collection. No synovitis, or synovial enhancements. Numerous nonspecific ill defined subcutaneous soft tissue lesions.
Additional Evaluation

- Echocardiogram is normal
- EKG: PR interval 120
- ASO and DNASE B antibody - negative

Rheumatic fever is ruled out

Differential Begins to Narrow

**Infectious**
- Septic joint
- Rheumatic Fever
- Lyme disease
- Osteomyelitis

**Rheumatologic**
- Reactive Arthritis
- Systemic JIA
- Rheumatic fever
- Lupus Vasculitis
- Kawasaki
- Granulomatous disease
- Sarcoidosis

**Heme/Onc**
- Lymphoma
- Histiocytosis

**Orthopedic**
- Fracture
- Dislocation
- Trauma
- Growing pains
- Legg-calve-perthes
- Avascular necrosis
Further Laboratory Work Up

**Differential was expanded**

- Angiotensin converting enzyme - normal
- Lysozyme - normal
- Vitamin D 25 hydroxy: 28.4 (L)
- Vitamin D 1,25 hydroxy: 84 (H normal is 79)
- Ferritin: 93
- FOBT: negative
- LD: 919 (H)
- Uric acid 3.0
- Skin biopsy: erythema nodosum
- Ophthalmologic exam: no uveitis
- ANCA <1:20 (Negative)
- RO and LA antibody normal
- Smith antibody normal
- ANA – negative
- C4 46
- C3 159

Chest X-ray

Ill-defined reticular/micronodular bibasilar opacities with probable hilar and mediastinal adenopathy. CT may be helpful for further evaluation.
CT Scan

Perilymphatic and random miliary nodules spread diffusely throughout the lungs. Bilateral posterior pleural thickening. There are bilateral hilar lymph nodes.

Work up is indicative of granulomatous disease

- Sarcoidosis is moving up on the differential but this is uncommon in children.
- Other infectious potential causes of symptoms which were explored
  - Histoplasmosis – negative
  - Tuberculosis – negative
  - Toxoplasmosis – negative
  - Bartonella - negative
What test diagnoses sarcoidosis?

1. Biopsy with non-caseating granuloma
2. Elevated 1-25 dihydroxyvitamin D
3. Hilar Lymphadenopathy on chest x-ray
4. Elevated angiotensin converting enzyme
5. No one test it is a diagnosis of exclusion

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Sarcoidosis

Cause and Diagnosis

- No definitive cause
  - One theory: susceptible individual plus environmental exposure causing
- Prevalence in adults is 10 in 100,000 and in children is 0.22-0.27 in 100,000
- Need compatible clinical findings with non-caseating granulomas with exclusion of other granulomatous disorders

Signs and Symptoms of Sarcoidosis

- Lungs – granulomas, fibrosis (hilar lymphadenopathy)
- Lymph nodes - (often good place to get a tissue sample)
- Eyes - uveitis, keratotic precipitates in cornea
- Skin - erythema nodosum or soft flat topped papules on the face and plaques on the trunk
- Joints – effusions, pain
- Kidney – proteinuria, hematuria
- Liver - cirrhosis or asymptomatic granulomas
- Fatigue, malaise, and fever
Work up of Sarcoidosis

Tests Which Can Indicate Sarcoidosis

- Labs
  - Hypercalcemia, elevated 1-25 dihydroxyvitamin D, elevated ESR and CRP, Elevated Angiotensin Converting Enzyme
- CXR
  - bilateral hilar lymphadenopathy
- CT scan
  - confirm bilateral hilar lymphadenopathy
- BAL
  - increased lymphocytes, CD4/CD8 ratio is high
- Tissue showing noncaseating granuloma

This Patient’s Presentation

<table>
<thead>
<tr>
<th>What fits</th>
<th>What does not fit</th>
</tr>
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<tbody>
<tr>
<td>Chest x-ray showing hilar lymphadenopathy</td>
<td>No documented noncaseating granuloma (skin biopsy showed erythema nodosum but no granulomas)</td>
</tr>
<tr>
<td>CT showing lymphadenopathy</td>
<td>No eye involvement (ophthalmology consulted for eye exam which was normal)</td>
</tr>
<tr>
<td>High 1,25 hydroxyvitamin D</td>
<td>No arthritis on exam</td>
</tr>
<tr>
<td>Erythema Nodosum</td>
<td>Normal Angiotensin Converting Enzyme</td>
</tr>
<tr>
<td>Elevated ESR and CRP</td>
<td></td>
</tr>
</tbody>
</table>
How is Sarcoidosis Treated?

1. Supportive Measures Only
2. NSAIDS
3. Steroids
4. Antibiotics
5. IVIG

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Treatment

Where is our patient now?

• Rheumatology was consulted
• Treatment is steroids 1-2 mg /kg/day for 1-2 months until symptoms resolve or improve
• Steroids are tapered slowly and clinical response is monitored closely
• Methotrexate can be used if steroid resistance is observed

Conclusion

• When you don’t know exactly what is happening always rule out life threatening things first
• Common things are common and patients don’t read the text book
• Sarcoid is rare and is a diagnosis of exclusion
• Because the treatment of sarcoidosis is steroids, providers need to rule out other infectious processes before starting therapy
References


Thank You

Dr. Ashford
Dr. Reinhardt