

A 6-year-old girl is brought to the physician with a rash and joint pains. She had previously been healthy except for a sore throat a few weeks ago that resolved on its own. Last week, the patient developed pain in her knees. The pain resolved after a few days, but now her ankles and wrists are tender. She has also developed a non-pruritic pink rash on her back. Temperature is 38.3 C (101 F), pulse is 85/min, and respirations are 20/min. On examination, there is pain and stiffness with manipulation of the wrists and ankles. A **faint, erythematous, centrifugal rash** is present on her trunk and proximal limbs. Lungs are clear and no murmur is heard. Serum laboratory results are as follows:

Leukocytes	6,500/ μ L
Neutrophils	56%
Lymphocytes	33%
Hemoglobin	12.5 g/dL
Platelets	200,000/ μ L
C-reactive protein	35 mg/dL
Erythrocyte sedimentation rate	38 mm/hr

Which of the following is the most likely diagnosis?

- ☐ A. Acute lymphoblastic leukemia
- ☐ B. Acute rheumatic fever
- ☐ C. Juvenile idiopathic arthritis
- ☐ D. Henoch-Schönlein purpura
- ☐ E. Systemic lupus erythematosus

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Media Exhibit

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Which of the following is the most likely diagnosis?

- ☐ A. Acute lymphoblastic leukemia [1%]
- ☒ B. **Acute rheumatic fever** [67%]
- ☐ C. Juvenile idiopathic arthritis [23%]
- ☐ D. Henoch-Schönlein purpura [8%]
- ☐ E. Systemic lupus erythematosus [2%]

Proceed to Next Item

Explanation:

User Id:

Acute rheumatic fever

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Explanation:

User Id: [REDACTED]

Acute rheumatic fever		
Epidemiology	<ul style="list-style-type: none">• Peak incidence: Age 5-15• Twice as common in girls	
Clinical features	Major	<ul style="list-style-type: none">• Joints (migratory arthritis)• ♥ (Carditis)• Nodules (subcutaneous)• Erythema marginatum• Sydenham chorea
	Minor	<ul style="list-style-type: none">• Fever• Arthralgias• Elevated erythrocyte sedimentation rate/ C-reactive protein• Prolonged PR interval
Late sequelae	Mitral regurgitation/stenosis	
Prevention	Penicillin for group A streptococcal (<i>Streptococcus pyogenes</i>) pharyngitis	

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Acute rheumatic fever (ARF) is a complication of untreated group A streptococcal pharyngitis, which usually precedes the onset of rheumatic fever by 2-4 weeks. ARF is diagnosed clinically using the Jones criteria, which are split into major and minor criteria (Table). The diagnosis is based on evidence of a preceding group A streptococcal infection along with 2 major criteria, or 1 major plus 2 minor criteria.

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This patient's **polyarthrititis, erythema marginatum** (an evanescent pink rash with sharp edges), fever, and elevated acute phase reactants (C-reactive protein [CRP] and erythrocyte sedimentation rate [ESR]) satisfy the diagnostic criteria for ARF. Supportive laboratory findings include a positive streptococcal antigen test or elevated antistreptolysin O titer. Rheumatic fever can be prevented with penicillin treatment of streptococcal pharyngitis.

(Choice A) Acute lymphoblastic leukemia is a malignancy of white blood cells that causes lymphocytosis or lymphopenia along with anemia or thrombocytopenia. This patient has a normal complete blood count.

(Choice C) Juvenile idiopathic arthritis is diagnosed when arthritis is present for >6 weeks. Systemic symptoms, including a rash, can be seen in children with juvenile idiopathic arthritis, but the arthritis is usually not migratory, as in this patient.

(Choice D) Henoch-Schönlein purpura is a vasculitis that can cause a transient, migratory arthritis and rash. However, the rash is purpuric and the arthritis typically involves the lower-extremity joints (hip/knee/ankle).

(Choice E) Systemic lupus erythematosus is an autoimmune disease that affects multiple organ systems. Arthritis and elevated CRP and ESR can be seen, but the rash is usually present across the cheeks (malar rash). Multiple organ system involvement (eg, hematologic, neurologic, renal) is needed for the diagnosis.

Educational objective:

Rheumatic fever is a preventable complication of streptococcal pharyngitis. It is diagnosed clinically using the Jones criteria. The major criteria include carditis, migratory polyarthrititis, Sydenham chorea, subcutaneous nodules, and erythema marginatum.

References:

1. [Acute rheumatic fever and streptococci: the quintessential pathogenic trigger of autoimmunity.](#)
2. [Diagnostic criteria of acute rheumatic fever.](#)
3. [Clinical characteristics and cardiac outcome of acute rheumatic fever in](#)

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2. [Diagnostic criteria of acute rheumatic fever.](#)
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