

A 9-month-old African American boy is brought to the office by his parents due to swelling of the feet and hands for the past two days, accompanied by poor feeding and fussiness. His vital signs are stable, except for a temperature of 38.3° C (101° F). He appears pale. On examination, the dorsal surface of his hands and feet on both sides are swollen and tender, with restricted range of movement. He is an adopted child, and his family history is not available. Radiography of the feet and hands reveal soft tissue swelling. What is the pathophysiology of this patient's presentation?

- ☐ A. Salmonella osteomyelitis
- ☐ B. Vaso-occlusive phenomena
- ☐ C. Hypertrophic osteoarthropathy
- ☐ D. Staphylococcus osteomyelitis
- ☐ E. Autoimmune phenomena



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- ☐ A. Salmonella osteomyelitis [7%]
- ☒ B. Vaso-occlusive phenomena [78%]
- ☐ C. Hypertrophic osteoarthropathy [2%]
- ☐ D. Staphylococcus osteomyelitis [3%]
- ☐ E. Autoimmune phenomena [9%]

[Proceed to Next Item](#)

### Explanation:

User Id: XXXXXXXXXX

Hand-foot syndrome or dactylitis is the earliest manifestation of vaso-occlusion in sickle cell anemia, thereby warranting a complete workup for previously asymptomatic sickle cell patients. Patients usually present at the age of 6 months to 2 years with an acute onset of pain and symmetric swelling of the hands and feet. Although only soft tissue swelling is initially seen, fever is sometimes present. The pathophysiology of dactylitis involves vascular necrosis of the metacarpals and metatarsals, which may be seen on plain radiographs as osteolytic lesions.

**(Choice A)** Patients with sickle cell disease have poor splenic function, and are thus more prone to develop septic joints and osteomyelitis. In such patients, osteomyelitis usually involves single bones (rather than bilateral hands and feet), and *Salmonella* is often isolated from bone cultures.

**(Choice C)** Hypertrophic osteoarthropathy is a clinical syndrome comprised of clubbing of the toes and fingers, swollen joints and enlarged extremities. It may be primary or secondary. The primary form is very rare. The secondary form occurs in patients with pulmonary diseases, usually cancer. Lung cancer is very unlikely at this age group.

**(Choice D)** Staphylococcal osteomyelitis presents with localized bone pain, soft tissue swelling and erythema. Sinus tracts with purulent drainage can sometimes be found on the skin surface. X-rays are initially normal, but will later show (after approximately two to



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**(Choice D)** Staphylococcal osteomyelitis presents with localized bone pain, soft tissue swelling and erythema. Sinus tracts with purulent drainage can sometimes be found on the skin surface. X-rays are initially normal, but will later show (after approximately two to three weeks) periosteal elevation and necrotic areas. In sickle cell patients, *Salmonella* often causes osteomyelitis.

**(Choice E)** Sickle cell disease is not an autoimmune disorder. Its pathology involves a point mutation which results in the substitution of valine for glutamic acid on the surface of the Hb S molecule in the sixth codon of the beta globin chain.

### Educational Objective:

Dactylitis is the earliest manifestation of vaso-occlusive disease in sickle cell anemia.

Time Spent: 20 seconds

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Last updated: [08/15/2016]