

History

A four month-old boy presented with abdominal distension. His parents had noted increasing abdominal size over several weeks. They also reported a diaper rash for two months and nasal congestion for one month. They denied fever, cough, emesis, or diarrhea. The patient was adopted, born full term in another state. Birth mother had limited prenatal care.

Notable Exam Findings

HEENT: Nasal congestion without discharge.

ABDOMEN: Distended, with spleen palpable to the level of the umbilicus and liver 5cm below the costal margin.

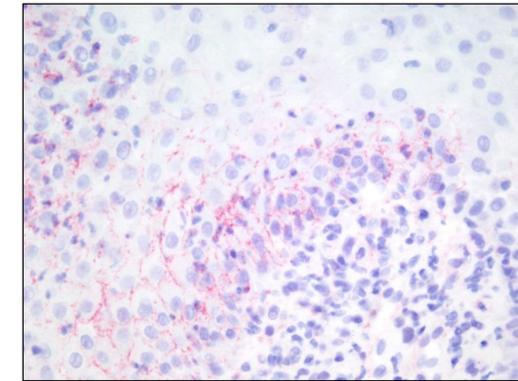
SKIN: An erythematous, macular diaper rash in the perineum with several 0.5 cm macules on the inside of both thighs.

NEURO: Alert and interactive, mild hypotonia.

Laboratory Findings

- Hemoglobin - 7.8
- AST 101, ALT 56
- Serum chemistry, Uric Acid, LD, Total Bilirubin, CEA, and BHCG - Normal
- HIV, HSV, EBV and CMV testing: Negative
- **Serum RPR - 1:8192**
- **CSF VDRL - Reactive**

Photos and Pathology



From Left: Photo showing hepatosplenomegaly, with liver and spleen edges demarcated. Photo of erythematous macular diaper rash. Photo of biopsy of rash showing spirochetes.

Radiographic Images



From Left: X- Rays showing medial metaphaseal destruction of both tibias (Wimberger's sign, superior arrows Panels A, B and C) and diffuse tibial periosteal reactions (inferior arrows, Panels A, B and C). Far Right: MRI showing hepatomegaly and splenomegaly.

References and Acknowledgements

1. US Center for Disease Control 2012 Sexually Transmitted Diseases Surveillance, <http://www.cdc.gov/std/stats12/tables/1.htm>, accession Jan 12, 2014
2. Screening for Syphilis Infection in Pregnancy: U.S. Preventive Services Task Force Reaffirmation Recommendation Statement. *Ann Intern Med.* 2009;150:705-709.
3. Berkowitz K, Baxi L, Fox HE. False-negative syphilis screening: The prozone phenomenon, non-immune hydrops, and the diagnosis of syphilis during pregnancy. *Am J Obstet Gynecol.* 1990;163: 975-977.
4. US Center for Disease Control Sexually Transmitted Disease Treatment Guidelines 2006, Congenital Syphilis, <http://www.cdc.gov/std/treatment/2006/congenital-syphilis.htm>, accession Jan 12, 2014

Discussion

Congenital syphilis is uncommon in the United States, with an incidence of 7.8 cases/100,000 live births. Congenital syphilis may cause stillbirth, hydrops fetalis or prematurity, but the clinical presentation can vary greatly and no signs may be apparent at birth. Classic signs and symptoms in infants include hepatosplenomegaly, rash, "snuffles," jaundice, pseudoparalysis, transaminitis, and anemia.

Diagnosis is made with serum RPR. The USPSTF recommends prenatal RPR screening. False negatives have been described in pregnant mothers with very high titers, the so-called "prozone effect." In this case, the patient was not screened. Darkfield microscopy may be performed on any nasal discharge or suspicious skin lesions.

Conclusion

Our case demonstrates the need for strict adherence to evidence-based protocols for syphilis screening during pregnancy and delivery. For the general pediatrician, maintaining a good knowledge base of the protean manifestations of congenital syphilis will be invaluable in prompt diagnosis and treatment of this old foe.