CJIM | Clinical Science

Parrot's Pseudoparalysis in Congenital Syphilis

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Introduction: Congenital syphilis is an intrauterine infection transmitted by the spirochete Treponema pallidum, and it is the most common congenital infection in the world. Cases of congenital syphilis continue to rise in the United States, and prompt clinical diagnosis of those cases that escape prenatal screening is critical given the wide availability of treatment and prevention of long-term sequela when treatment is provided.

Case Presentation: Here we report an infant who presented with irritability and apparent paralysis of the right upper extremity, consistent with Parrot's pseudoparalysis, a classic physical exam finding in congenital syphilis. The infant was treated with intravenous penicillin G for 10 days. In the following months, the infant's symptoms improved – she was able to move all limbs comfortably and the pseudoparalysis completely subsided. Cranial and eye exams were normal. Her liver function tests normalized, and repeat non-treponemal antibodies fell at a slow rate over several months. Her growth and development were appropriate for age.

Conclusions: This case illustrates the importance of recognizing pseudoparalysis of Parrot and of considering congenital syphilis in an infant with bone pain or apparent paresis. Prior reports of pseudoparalysis of Parrot are reviewed.

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Introduction

Congenital syphilis is an infection passed transplacentally from mother to child and caused by the spirochete, Treponema.[1] Infants with congenital syphilis can present with devastating symptoms affecting any organ system. Skeletal manifestations are common in syphilis infection, and, in rare cases, bone involvement may even be the presenting symptom.[2] Parrot's pseudoparalysis is a physical exam finding in which painful periostitis results in decreased or absent movement of an extremity secondary to pain.[2] Prompt recognition of Parrot's pseudoparalysis may prevent radiation and sedation exposure.

Syphilis is the most common congenital infection in the world, and it has the potential to impose significant consequences when left untreated.[3] Congenital syphilis cases in the United States have increased 261% from 2013-2018;[4] though once considered a rare diagnosis, escalating numbers suggest a need for clinicians to keep congenital syphilis in mind. In fact, new cases are reported in regions where infection was once uncommon, such as the United Kingdom.[5]

Among barriers to early diagnosis, inadequate maternal treatment and lack of timely prenatal care are the most common.[4] While robust prenatal and perinatal screening have the potential to identify most infants at risk, it is imperative to recognize symptoms in those who are missed by screening. A comprehensive understanding of the skeletal manifestations of congenital syphilis may lead to prompt diagnosis and treatment, thus decreasing morbidity and mortality. Here we describe a case of an adopted infant with limited prenatal care who presented with isolated pseudoparalysis of

Parrot.

Clinical Presentation

A two-month old ex-full term infant female presented to the emergency department from her pediatrician's office with irritability for one week and refusal to move her right upper extremity. The patient's medical history was notable for a two-week hospital stay after birth for treatment of neonatal abstinence syndrome secondary to maternal methamphetamine use. She was discharged home with her adopted parents. At two months old, the patient presented to her pediatrician with irritability for one week and paucity of movement in her right upper extremity for several hours. When dressing the child for the office visit, she appeared to be moving her left upper extremity normally but was not moving her right upper extremity. It was unclear whether her decreased movement was related to pain or nerve conduction. She was sent to the emergency department for further evaluation.

On examination in the emergency department, the patient had spontaneous movement of all extremities other than her right upper extremity. No spontaneous movement of the right arm or forearm was noted; however, she did have an intact grasp reflex and movement of the right hand, suggesting that nerve conduction pathways were intact. There was tenderness to palpation at the right elbow and upper extremity, with subtle edematous soft tissue swelling around the right elbow joint. She appeared uncomfortable and cried with any passive movement of the right upper extremity. Laboratory evaluation was significant for transaminitis (AST 440; ALT 275), anemia (Hemoglobin 8.6), and elevated C-Reactive Protein (2.29) and erythrocyte sedimentation rate (45). Directed radiographs of the right upper extremity and elbow were unrevealing (**Figures 1 and 2**).

Concern for infectious etiology prompted testing for congenital infections. Rapid Plasma Reagin results returned reactive with a titer of 1:64, confirming a diagnosis of congenital syphilis. Venereal Disease Research Laboratory (VDRL) test was non-reactive. Blood culture and smear were unrevealing. Skeletal survey showed no metaphyseal or bony changes, and no acute fracture or dislocation. A lumbar puncture was performed to rule out neurosyphilis infection, and results were unremarkable. There was no evidence of ophthalmic findings including keratitis, uveitis, cataracts, glaucoma, retinitis, or optic neuropathy. Orthopedic surgery was consulted and no further arthropathy was detected.

The patient was placed on 50,000 units of intravenous penicillin G per kilogram of body weight for 10 days. After a second dose of treatment with penicillin, the infant developed a diffuse, flat, maculopapular rash that extended from the trunk to the bottom of her feet (**Figure 3**). The rash disappeared several hours later. She continued to receive penicillin G for a 10-day course, and her right arm pain and fussiness subsided. She was cleared medically and sent home after a 10-day treatment.

The patient had a few healthcare visits in the month prior to her presentation at the emergency department. At the Special Infant Care Clinic, she was noted to be well appearing, developing normally, and using both arms equally. Silver nitrate was applied to her umbilical granuloma at the visit. A few days later when she saw her primary care provider, where the patient was noted to be a little fussy, silver nitrate was again applied to her umbilical granuloma.

In the months following diagnosis of congenital syphilis and appropriate treatment, the infant's symptoms significantly improved. She was able to move all limbs, as the limb pain and paresis completely resolved. Her cranial and eye exams were normal. Her liver function tests went down to normal levels and repeat non-treponemal antibodies fell at a slow rate over several months. Her growth and development milestones were appropriate for her age.

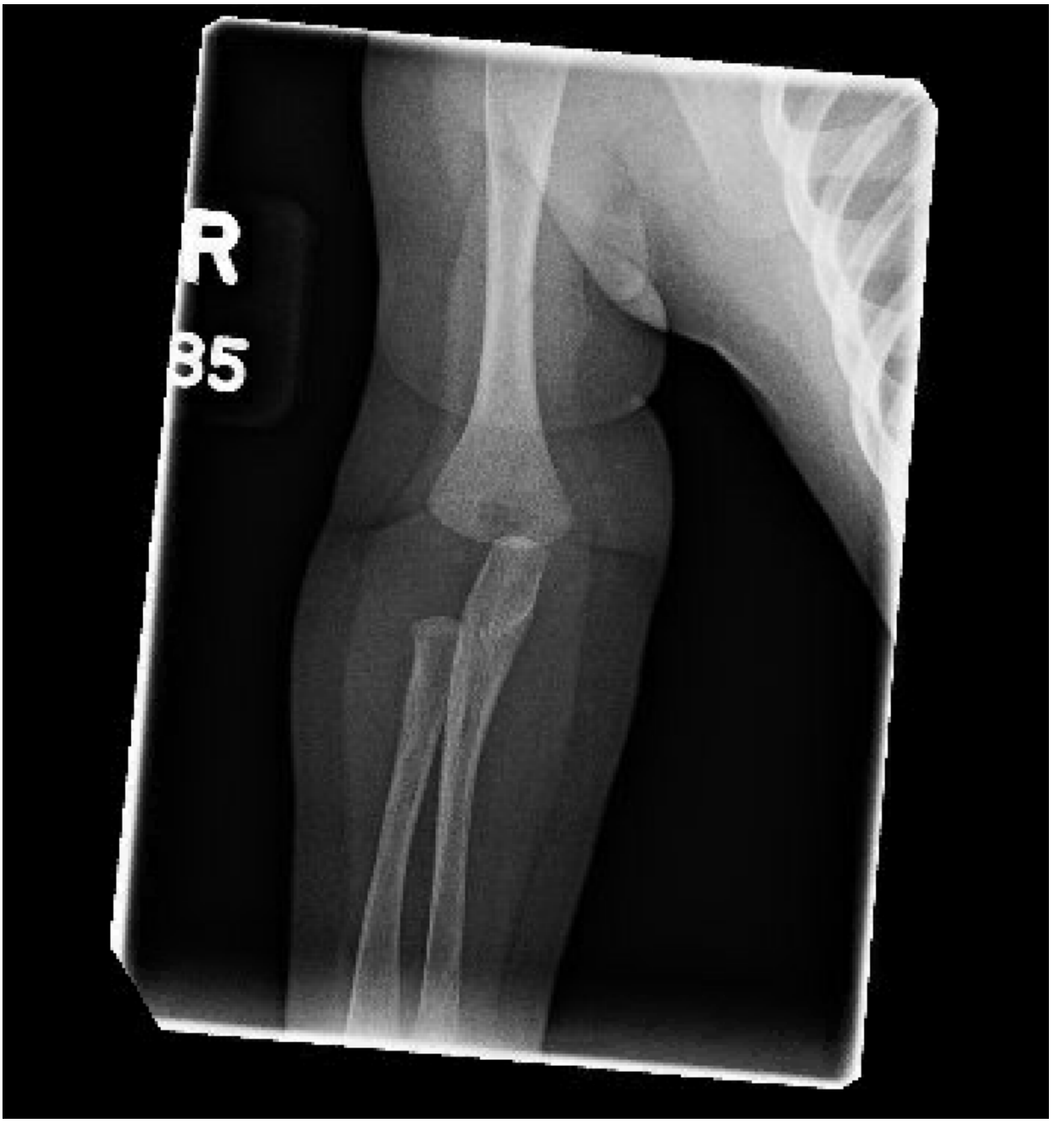

Figure 1. Radiographs of the right upper extremity (Figure 1) and right elbow (Figure 2), made when the infant was 2 months old, demonstrating no irregularities. A radiograph of the right upper extremity revealed subtle cortical irregularity involving the proximal radius and ulna, suggesting a projectional or non-displaced fracture. These radiographic findings suggested no notable osseous abnormalities.

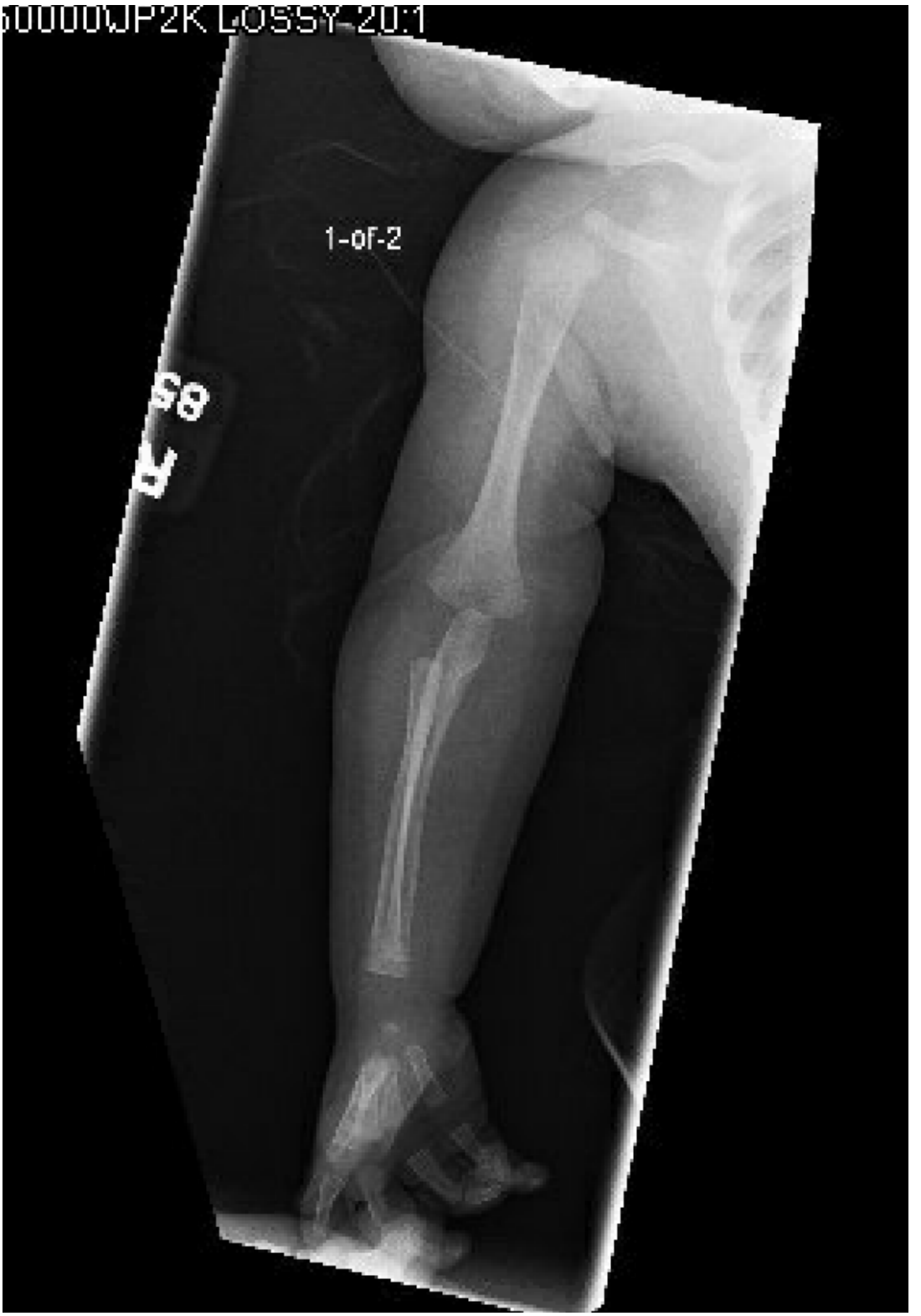

Figure 2. Radiographs of the right upper extremity (Figure 1) and right elbow (Figure 2), made when the infant was 2 months old, demonstrating no irregularities. A radiograph of the right upper extremity revealed subtle cortical irregularity involving the proximal radius and ulna, suggesting a projectional or non-displaced fracture. These radiographic findings suggested no notable osseous abnormalities.

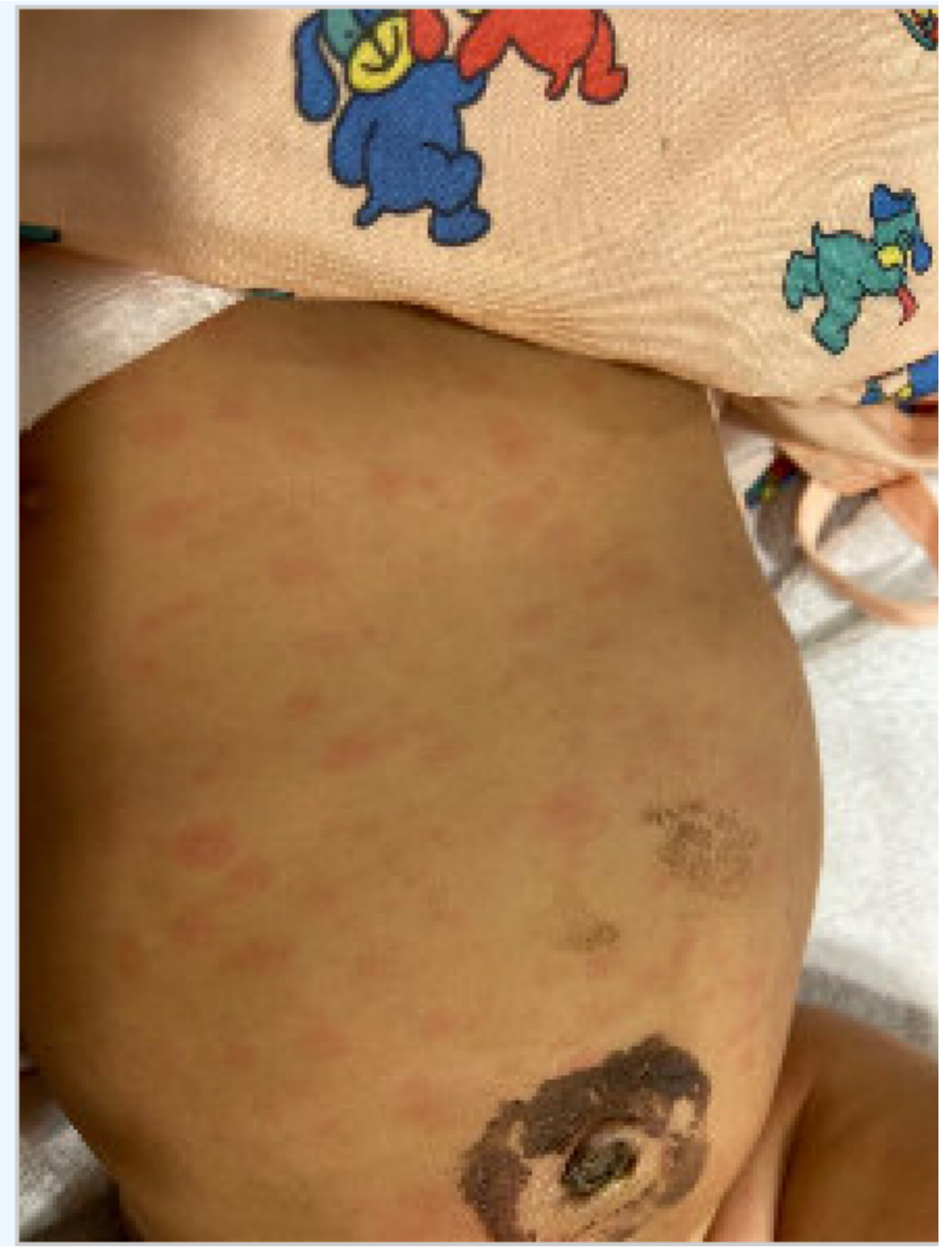

Figure 3. A diffuse, flat, maculopapular rash developed on the infant's trunk hours after administration of intravenous penicillin G, consistent with the Jarisch-Herxheimer reaction.

Discussion and Conclusion

The diagnosis of congenital syphilis based on symptoms alone can be particularly challenging. Infants may present with classic symptoms such as hepatosplenomegaly, snuffles, maculopapular skin rash, anemia and jaundice; they could also be asymptomatic.[6] Skeletal manifestation of congenital syphilis were first described by the French pediatrician Joseph Marie Jules Parrot and are thus termed pseudoparalysis of Parrot.[7,8] Pseudoparalysis of Parrot is characterized by apparent paralysis, particularly of the upper limbs, which is secondary to pain caused by inflammation around the bone.[9,10] Involvement of the spirochete bacterium is particularly evident where the bone is proliferating and is frequently located in the metaphasis of long bones.[2] Pseudoparalysis of Parrot can be intensely painful to the infant, so the infant will prefer to not move the bone at all. In this case, the infant presumably presented with this notable symptom of congenital syphilis. However, unilateral upper extremity pain with movement is not a typical presentation of pseudoparalysis.

Within the first 24 hours of penicillin G administration, the infant developed a diffuse maculopapular rash that is most characteristic of the Jarisch-Herxheimer reaction. When penicillin is administered in the setting of Treponema infection, the breakdown of spirochetes leads to an inflammatory response with fever, shaking chills, and diffuse rash. The Jasrish-Herxheimer reaction is caused by the release of lipoproteins and non-endotoxin pyrogen from the spirochetes in response to treatment.[11] The Jarisch-Herxheimer reaction requires supportive care with a favorable prognosis. Eruption of this maculopapular rash further supports a syphilis diagnosis.

Radiographic workup in our case was unremarkable, which is unusual in symptomatic congenital syphilis as up to 95% of patients with involvement of the metaphysis of long bones have radiographic alterations.[10] Radiographic abnormalities can also be noted in 20% of asymptomatic congenital syphilis infections.[12] When a diagnosis of congenital syphilis is suspected, the Centers for Disease Control recommends radiographs of the infant's long bones.[13] Our case suggests that benign radiographic findings do not always rule out a diagnosis of syphilis infection.

Congenital syphilis has a variable presentation, and bone involvement occurs in 60-80% of all cases of early congenital syphilis.[12] Bone involvement in non-accidental trauma should be considered in an infant who presents with unilateral arm weakness and pain with no clear history of injury. In this case, however, early recognition of congenital syphilis prevented an extensive and potentially harmful workup. By bearing the atypical presentation of congenital syphilis in mind, a provider can more succinctly distinguish between different potential diagnoses and prevent delays in management.

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