



**Pediatric Imaging** Case Report

## Congenital syphilis, the great mimicker, confused with non-accidental trauma

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### ABSTRACT

Congenital syphilis (CS) remains a major health problem worldwide. Children born with CS can present with variable symptoms that can mimic other disease processes. This report describes the case of a 4-month-old infant who presented with the left wrist swelling and distal radial fracture, causing concern for non-accidental trauma (NAT). An osseous survey showed additional findings and led to the correct diagnosis. Although radiographic features of NAT may overlap with CS, certain features such as bilaterally symmetrical skeletal involvement and metaphyseal destruction, as seen in our case, can help distinguish between the two.

**Keywords:** Congenital syphilis, Syphilis, Non-accidental trauma

### INTRODUCTION

Congenital syphilis (CS) is an ancient disease that continues to be a major health problem despite optimal preventive measures. The incidence and geographical spread of CS have increased in the United States over the last decade.<sup>[1,2]</sup> CS can affect various organ systems including the liver, nervous system, and bones. Children born with CS can present with signs and symptoms that can mimic many diseases. Skeletal changes may be the presenting symptom as in our case. Skeletal changes caused by CS include periostitis, metaphyseal destruction, and insufficiency fractures. Metaphyseal fractures in CS can mimic non-accidental trauma (NAT) and the two can be differentiated by thorough clinical examination, imaging, and laboratory tests.

### CASE REPORT

A 4-month-old girl presented to her pediatrician's office for a routine checkup and was noted to have left wrist swelling and deformity. Radiographs showed a healing angulated fracture of the left distal radius [Figure 1]. Given the patient's age and unexplained mechanism of injury, concern for NAT was raised. She was called back to the emergency department. Her mother denied any history of injury, sick contacts, or travel. The patient was born at 41 weeks with a birth weight of 3.22 kg. Maternal rapid plasma reagin (RPR), human immune deficiency virus, and hepatitis B tests were negative during her pregnancy.

On the physical examination, the patient appeared well, with no fever, fussiness, or wrist tenderness. The patient was afebrile (temperature 37°C). Laboratory analysis showed a white blood cell count of  $27.5 \times 10^9/L$ , hemoglobin of 7.7 g/dL with microcytosis, C-reactive



**Figure 1:** A 4-month-old girl presented with the left wrist swelling. The left wrist radiograph shows a distal radial metaphyseal transverse fracture (black arrow) and metaphyseal lucency involving the distal ulna (double-sided black arrow). Periostitis involving both radius and ulna is also seen.

protein of 32 mg/L, and erythrocyte sedimentation rate of 106 mm/h.

An osseous survey was ordered as part of the workup for suspected NAT. In addition to bilateral distal radial metaphyseal fractures, the osseous survey revealed periostitis involving long bones of both upper and lower extremities [Figures 2 and 3] and metaphyseal destruction of several bones with the most pronounced involvement of bilateral proximal medial tibial metaphyses, Wimberger sign [Figures 4 and 5]. Based on the radiographic findings, concern for CS was raised. The skeletal survey did not show any rib fractures, metaphyseal corner fractures, or other fractures specific to NAT. Ultrasound of the abdomen showed splenomegaly but was otherwise normal. A computed tomography scan of the head was normal, and an ophthalmological examination did not show evidence of retinal hemorrhages. The diagnosis of CS was confirmed with a positive rapid plasma reagin (RPR, 1:128 titer), *Treponema pallidum* IgM antibody, and IgG antibody. The infant was admitted to the hospital and treated with 10 days of intravenous penicillin. The mother admitted to unprotected intercourse following the last set of laboratory tests during her pregnancy. Maternal RPR was repeated and came back to be positive. She was treated for syphilis.

## DISCUSSION

CS occurs because of vertical transmission of infection from an infected mother to the baby. It can lead to spontaneous abortion, stillbirth, premature birth, or low birth weight.



**Figure 2:** A 4-month-old girl presented with the left wrist swelling. The right forearm radiograph shows metaphyseal destruction involving the distal humerus (double-sided black arrow) and distal ulna and transverse distal radial metaphyseal fracture. Periostitis involving the humerus, radius, and ulna is also seen.

The incidence of CS in the United States has increased significantly in the last decade. There was a 477% increase in the reported incidence of CS in 2019 relative to 2012 (CDC.gov). Controlling community transmission and identification and treatment of pregnant women are important to decrease the incidence and infant morbidity and mortality due to CS. The Centers for Disease Control and Prevention, American Academy of Pediatrics, US Preventive Services Task Force, and American College of Obstetricians and Gynecologists recommend first-trimester screening for all pregnant women and additional screening in the third trimester and at the time of delivery for women who are at increased risk.

Clinical manifestations of syphilis are divided into early (occurring within the first 2 years of life) and late (occurring within the first two decades of life).<sup>[3]</sup> Common early clinical manifestations include rhinitis and snuffles, rash, fever, failure to thrive, pseudo paralysis, hepatosplenomegaly, jaundice, lymphadenopathy, pneumonitis, central nervous system involvement, and ascites.<sup>[3]</sup> Long-term outcomes of CS include developmental delay, microcephaly, and growth restriction. Laboratory analysis shows leukocytosis, anemia, thrombocytopenia, proteinuria, and hematuria. Skeletal changes are seen in 60–80% of symptomatic infants.

**Table 1: Specificity of fractures for non-accidental trauma.**

High specificity	Moderate specificity	Low specificity
Classic metaphyseal lesions/metaphyseal corner fractures/bucket-handle fractures	Multiple fractures	Fractures of shafts of long-bones
Multiple rib fractures (posterior or posteromedial)	Fractures of different ages	Clavicular fractures
Scapular fractures	Digital fractures	Linear skull fractures
Spinous process fractures	Complex skull fractures	
Sternal fractures	Vertebral body fractures	



**Figure 3:** A 4-month-old girl presented with the left wrist swelling. The right femur radiograph shows periostitis and distal femoral metaphyseal destruction (double-sided black arrow).

Radiographic findings of CS include diffuse periosteal reaction, metaphyseal demineralization, and irregularities. Metaphyseal destruction of the upper medial tibial metaphysis is called the “Wimberger sign.” Insufficiency fractures through the weakened metaphysis may be seen.<sup>[4]</sup> The lower limb bones are most affected.<sup>[4]</sup> Bilaterally symmetrical involvement is characteristic of CS. Radiographic findings of CS can be confused with NAT.<sup>[5,6]</sup> Skeletal manifestations of NAT include multiple fractures involving different bones and at various stages of healing. Certain fractures are more specific for NAT such as posterior rib fractures, metaphyseal corner fractures, fractures of the scapula, spinous processes, and sternum [Table 1].<sup>[7]</sup> Exuberant callus and periosteal bone formation can also be seen in NAT. Although fracture in a non-ambulatory child should always prompt concern for NAT



**Figure 4:** A 4-month-old girl presented with the left wrist swelling. The left leg radiograph shows proximal medial tibial metaphyseal destruction (Wimberger sign) (double black arrows), and distal fibular and distal tibial metaphyseal destruction.

and any fracture in a child can be due to intentional trauma, other differential diagnoses should be considered. Classic metaphyseal fractures of child abuse have bucket handle or corner fracture morphology, due to the specific mechanism of injury, that is, pulling and twisting of the limb or flailing of an unsupported limb.<sup>[7]</sup> The distal radial metaphyseal fractures in our case did not have a bucket-handle morphology. The skeletal survey in our patient did not show posterior rib fractures or other high specificity fractures, such as fractures of the scapula or spine. Furthermore, classic findings of CS, such as bilateral periostitis and metaphyseal destruction, were seen in the skeletal survey.

High clinical suspicion and correlation with history, mechanism of trauma, and developmental stage of the child



**Figure 5:** A 4-month-old girl presented with the left wrist swelling. The right leg radiograph shows proximal medial tibial metaphyseal destruction (Wimberger sign) (double black arrows) and distal tibial and fibular metaphyseal destruction.

need to be considered to identify cases of child abuse. It is equally important to consider other differential diagnoses in these clinical scenarios to ensure appropriate management of the child. When characteristic radiographic findings of bilateral periostitis and metaphysitis including the Wimberger sign are present, the diagnosis of CS can be made confidently.

## CONCLUSION

CS is called the great mimicker because it can present with various signs and symptoms and be confused with other diseases. Skeletal changes of CS include metaphyseal

destruction, periosteal reaction, and insufficiency fractures. Bilaterally symmetric bony changes are characteristic of CS. Radiologists should be aware of the skeletal findings seen in CS. Although a long bone fracture in an infant should raise concern for NAT, other differential diagnoses, including CS, should be considered. A skeletal survey can help make the correct diagnosis and laboratory tests can further help to confirm the diagnosis.

## Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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## Conflicts of interest

There are no conflicts of interest.

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