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CASE REPORT

Congenital Syphilis in a Patient with Bone Fractures: Case Report

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Abstract

Congenital Syphilis (CS) is an infection by Treponema pallidum; it is transmitted transplacentally or in childbirth. The disease manifests itself from asymptomatic to severe forms, including fetal and neonatal death. With a diverse clinical manifestation, it has many diagnostic possibilities when the diagnosis is unknown. This report aim to show a case of an infant with undiagnosed CS at birth, whose initial presentation was considered serious, with consecutive bone fractures, which led the medical team to consider, among the diagnostic hypotheses, the possibility of abuse; and also aims to discuss possible causes of therapeutic failure that culminated in CS.

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INTRODUCTION

Congenital Syphilis (CS) is caused by transmission of Treponema pallidum from mother to fetus via the placenta or at birth[1]. Epidemiological data show an increase in the number of cases of this notifiable disease in Brazil[2]. CS can be prevented as long as gestational syphilis (GS) is diagnosed prenatally and treated promptly[3].

The myriad possible clinical manifestations of the disease, ranging asymptomatic cases to fetal or neonatal death, increase the list of conditions that have to be ruled out in differential diagnosis until CS is confirmed. Treatment with penicillin is recommended and patients must be monitored until they are cured or diagnosed with sequelae from the disease[4-7]. This report describes an unusual case of CS that made the care team consider the possibility of abuse, and discusses the clinical findings and possible causes of failure in the treatment prescribed to the mother of the infant. The Ethics Committee of the institution at which the patient was provided care approved this article.

CASE REPORT

A male infant aged two months and eighteen days was brought to the emergency department of a pediatric hospital for crying whenever his right forearm was touched. Physical examination confirmed he was in pain whenever he moved his right arm. X-ray images showed a distal ulnar fracture. His mother claimed she was unaware of what caused the fracture. The infant's arm was placed in a cast.

Two days later, the infant was brought back to the emergency department with pain in the left arm. His arm was swollen and he felt pain on palpation. X-ray images showed a left distal ulnar fracture (Figure 1). His left arm was placed in a cast and an outpatient consultation was scheduled with an orthopedist.

Since the patient did not show up in the follow-up consultation, the hospital's social services were called to start a search on grounds of abuse and neglect.

A review performed in the patient's records revealed a history of GS, with treatment prescribed to the then pregnant mother and her partner. Serial non-treponemal tests (VDRL) performed on her showed titers of 1:256 (four weeks of gestation), 1:64 (18 weeks), 1:32 (31 weeks), and 1:16 (delivery).

The patient was born via vaginal delivery after 38 weeks and five days of gestation; he was 49.5 cm long, weighed 3410g, had a head circumference of 35 cm, and Apgar scores of 9/9. Neonatal screening tests (Ortolani, red reflex, hearing screening, pulse oximetry) were normal. The infant's VDRL titer at birth was 1:16, the same as his mother. A monitoring protocol was started to keep track of signs and symptoms and workup results. At eight weeks of age, his VDRL titer was 1:4, a drop of two dilutions.

He was admitted for further investigation. The infant had both arms in casts, diaper rash, nasal congestion, and whimpered when he was handled. His weight was plotted on a growth curve (weight x age), which yielded Z-scores between 0 and -2. Eye lesions and neurosyphilis were ruled out. X-ray images showed, in addition to the findings described above, lysis in the distal tibial metaphysis and bilateral diffuse periosteal reaction in the tibia and fibula (Figure 2). Abdominal ultrasound ruled out organomegaly. The VDRL titer was 1:512 and the patient had erythrocyte alterations, anemia, leukocytosis, thrombocytopenia, elevated C-reactive protein and alkaline phosphatase levels, clear cerebrospinal fluid with a negative VDRL test and a CSF protein content of 33 g/dL. Treatment with intravenous crystalline penicillin, 50,000 IU/kg/dose, every 6 hours, for ten days, was started.



Figure 1. Signs of fracture in the distal region of the ulna on both arms; x-rays of the arms of an infant diagnosed with congenital syphilis.

With the improvement in mobility after the start of treatment, the infant was diagnosed with pseudoparalysis of Parrot based on interview findings and physical examination. The patient's global development was appropriate for his age and he had no neurological or joint injuries.



Figure 2. Lysis in the distal metaphysis of the tibia and bilateral diffuse periosteal reaction in the tibia and fibula seen on X-ray images of the legs of an infant diagnosed with congenital syphilis.

The patient remained hospitalized for 16 days, as he improved gradually. In post-treatment outpatient follow-up, the VDRL titers at one, two and four months were 1:128; 1:32 and 1:8, respectively. Control X-ray images taken four months after treatment showed resolution of bone alterations. The patient remains under outpatient follow-up.

DISCUSSION

CS is an infectious disease caused by Treponema pallidum that affects multiple systems that can be transmitted via the placenta or at birth. Between 2009 and 2019, there were 6,600 miscarriages, 6,218 stillbirths and 1,835 deaths of children under one year of age due to CS in Brazil. The current incidence in the country is 8.2 cases per 1,000 live births[6]. According to Korenromp et al. (2019), the global number of cases of CS decreased slightly between 2012 and 2016, despite the stability in the prevalence of GS[8]. However, other studies described increases in the number of cases of CS. An increase of 261% in the number of cases of CS was reported in the United States between 2013 and 2018[9]. In England, incidence increased from 0.001 in 2015 to 0.014 cases per 1,000 live births in 2019[10].

In Brazil, there are specific guidelines concerning the prevention, diagnosis and treatment of CS, a notifiable disease.

The prevention of CS, implemented via prenatal screening in the first and third trimesters of pregnancy, aims to diagnose and treat pregnant women and their partners. Screening at birth and in the postpartum period aims to detect reinfection, treatment failure and infection in women who did not receive prenatal care or who became infected later during pregnancy. Prenatal care and monitoring of exposed children is particularly important in contexts of social vulnerability[4,5,7]. Prenatal screening, treatment and monitoring at birth and in the postpartum period, and partner adherence to treatment were deemed adequate in the case reported herein.

The diagnosis of CS is made based on signs of untreated or inadequately treated GS, the presentation of symptoms of disease, and microscopy or serology tests[3]. Many children are born without symptoms, particularly the ones born to mothers offered adequate treatment. Despite the protocols developed to treat these children, it is important to observe signs indicative of treatment failure, such as high VDRL values in the mother prior to treatment or in the newborn at birth, even when these values are the same as the ones observed in the mother. Investigation also includes complete blood counts, x-rays, cerebrospinal fluid testing and tests for vertically transmitted infections[1,4,5].

CS can be categorized as early or late. Early cases of the disease present in individuals of up to two years of age, and include findings such as prematurity, low birth weight, hematological changes, mucocutaneous lesions, low weight gain, meningitis, seizures, rhinitis, pseudoparalysis of Parrot and bone alterations, among others. Bone fractures are less frequent, but are a sign of severe involvement[11]. Late CS presents in individuals older than two years, and includes findings such as gum ulcers, paresthesia, and hearing loss, to name a few[1,2,4,5]. Our patient presented hematological changes, nasal congestion, bone fractures and bone alterations.

CS has a broad spectrum of clinical signs and symptoms, making it a diagnostic challenge. Depending on the context, abuse must be considered in differential diagnosis[11]. Our patient had a bone fracture in the first visit, returned with another fractured bone on the second visit and was not taken to the scheduled follow-up consultation. These factors combined led the team to consider the possibility of abuse and neglect, which prompted the involvement of the institution's social services. Jacobs et al. (2019) reported cases in which signs of CS were seen as consistent with abuse[11-13].

Treatment is performed with penicillin. Indications include confirmed or probable cases of CS; cases in which maternal treatment was inadequately performed; cases in which there is

no evidence showing that treatment was delivered; and cases in which treatment performed within less than four weeks from birth. Patients must be followed for two years or until non-treponemal tests are negative[1-5,14]. Our patient's mother was diagnosed during prenatal care, was adequately treated and presented decreasing VDRL titers. The newborn was tested at birth and during a childrearing visit and showed a reduction in VDRL titers from 1:16 (delivery) to 1:4 (eight weeks). Clinical signs and non-treponemal test performed on admission (VDRL 1:512) confirmed the diagnosis of infection.

The reasons for infection in newborns beg discussion. Studies have shown that failure occurs mainly when pregnant mothers are not adequately treated, drugs other than to penicillin are prescribed, treatment is not delivered based on the stage of disease or when prenatal care is flawed (poor adherence, unskilled healthcare workers, absence of screening and monitoring)[4,7]. The stage of the disease (early, latent or late) and the presence of other sexually transmitted infections (STIs) have also been associated with higher treatment failure rates[14]. When treatment is properly delivered, failure rates are very low. To date, there have been no reports of treponema resistance to penicillin[1,5]. Maternal reinfection occurs when the pregnant mother has multiple partners, engages in unsafe sexual behavior or her partner does not adhere to treatment[1,2,5,15]. In the present case, the pregnant mother did not have another STI, had a steady partner and both were treated. However, the stage of the disease was unknown at the time of diagnosis, a possible cause of transmission of syphilis to the unborn child.

CONCLUSION

This case report described a wide variety of clinical signs of infection, often with symptoms that are not easily identified as related to the disease, stressing the need to perform a thorough review of the patient's clinical history.

Improvements are needed in the process used to investigate potential cases of GS, including access to quality prenatal care, STI testing, adequate treatment and follow-up. Minimizing failures in prenatal care and monitoring exposed children are equally important. The consequences of GS are predictable, preventable and treatable. Therefore, the number of cases of CS can be reduced.

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