Case Report

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Congenital Syphilis Simulating Bone Neoplasm in 2-Month Old Infant – Case Report

Luciana Friedrich[1]*, Júlia Lima Vieira[2], Cintia Junges[3], Veronica Indicatti Fiamenghi[3], Daniela Dias Morales[3] and Jiseh Loss[4]

[1]*Neonatology Section, Hospital de Clínicas de Porto Alegre.*

[2]*Federal University of Rio Grande do Sul - Brazil.*

[3]*Pediatrics Residence -Pediatric Department, Federal University of Rio Grande do Sul - Brazil.*

[4]*Pediatric Oncology Section, Hospital de Clínicas de Porto Alegre.*

***Corresponding author**
Luciana Friedrich, Rua Ramiro Barcelos, 2350-Hospital de Clínicas de Porto Alegre -11º Andar-Serviçode Neonatology, CEP 90035-903, Brazil, E-mail: lfriedrich@hcpa.edu.br.

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Abstract

Introduction: *Congenital Syphilis (CS) occurs through the transplacental transmission of Treponema pallidum in inadequately treated or non-treated pregnant women, and is capable of severe consequences such as miscarriage, preterm birth, congenital disease and/or neonatal death. CS has been showing an increasing incidence worldwide, with an increase of 208% from 2009 to 2015 in Brazil.*

Case report: *2-month old infant receives care in emergency service due to edema of right lower limb with pain in mobilization. X-ray with osteolytic lesion in distal fibula. Infant was sent to the Pediatrics Oncology clinic. Perinatal data: 7 prenatal appointments, negative serology at 10 and 30 weeks of gestation. End of pregnancy tests were not examined and tests for mother's hospital admission were not requested. Mother undergone elective cesarean section at 38 weeks without complications. During the pediatric oncologist appointment, patient showed erythematous-squamous lesions in neck and other scar-like lesions in upper body. A new X-ray of lower limbs showed lesions in right fibula with periosteal reaction associated with aggressive osteolytic lesion compromising distal diaphysis, with cortical bone rupture and signs of pathological fracture, suggestive of eosinophilic granuloma. She was hospitalized for a lesion biopsy. Laboratory tests: hematocrit: 23.1 / hemoglobin 7.7 / leukocytes 10,130 (without left deviation) / platelets 638,000 / Negative Cytomegalovirus IgG and IgM and Toxoplasmosis IgG and IgM / VDRL 1:128. Congenital syphilis diagnosis with skin lesions, bone alterations and anemia. Lumbar puncture: glucose 55 / total proteins 26 / VDRL non reagent / 13 leukocytes (8% neutrophils; 84% monomorphonuclear; 8% macrophages) and 160 erythrocytes / negative VDRL and culture. X-ray of other long bones, ophthalmological evaluation and abdominal ultrasound without alterations. Patient was hospitalized for 14 days for treatment with Ceftriaxone 100mg/kg/day, due to the lack of Crystalline Penicillin in the hospital. She is now under outpatient follow-up.*

Discussion: *CS is responsible for high rates of morbidity and mortality. The ongoing increase of cases of this pathology reflects a severe health issue and indicates failures in policies for the prevention of sexually transmitted diseases, with inadequate follow-up of prenatal and maternity protocols.*

Keywords: Congenital Syphilis, Infant, Prenatal.

Introduction

Syphilis is an systemic infectious-contagious disease, sexually transmittable and caused by Treponema pallidum, a Gram-negative bacteria of the Spirochete group. Congenital Syphilis (CS) occurs when the pregnant mother transmits the bacteria to the fetus via the placenta [1]. Prevention starts at prenatal care with periodic laboratory exams and follow-up until birth [2].

CS has been showing a growing prevalence in both developed and developing countries [2]. In 2015 in Brazil, 19,235 cases of CS were reported in children younger than 1 year old, with an incidence rate of 6.5 cases per 1,000 live births, totaling an increase of 20.4% in relation to 2014. In Rio Grande do Sul, the data is even more startling with 1,642 cases in 2015 and an incidence of 11.5 cases per 1,000 live births [3]. It is believed that the main factors associated with the increase in CS cases would be the poor preventative measures by the authorities and health agents, sexual precocity and promiscuity, an increase in the number of single and adolescent mothers, an unawareness by the population about the severity of the disease, AIDS, drugs use and abuse, and the inadequacy of prenatal care [2]. The consequences of CS can be severe, such as: miscarriage, preterm birth, various congenital

manifestations and even death of the newborn (NB) [1].

The presentation of CS can be divided into early and late. The early form appears until the second year of life and its main clinical manifestations are preterm birth, low birth weight, hepatosplenomegaly or splenomegaly, skin lesions such as palmoplantar pemphigus and flat condyloma, pneumonia, bloody nasal discharge, jaundice and general lymphadenopathy. Laboratory abnormalities include anemia, thrombocytopenia, leukocytosis and/or leucopenia [1]. Bone lesions are the most frequent manifestations, occurring in 70 to 100% of the cases, with pain to the active or passive movement of the limbs, irritability and immobility (pseudoparalysis) of the limbs observed. The main radiological alterations are periostitis, osteitis or osteochondritis [4].

The diagnosis must be established through epidemiological, clinical, laboratory and imaging criteria. Over half of the NBs are asymptomatic and, when present, the manifestations and symptoms are discreet or nonspecific, making the diagnosis complex [1]. All NBs of mothers diagnosed with syphilis during pregnancy or at birth, or in the clinical suspicion of congenital syphilis, must have specific investigation, including the NBs of properly treated mothers. The investigation is based on the completion of serum VDRL and, when indicated, supplemented with blood count, long bone X-rays and lumbar puncture with VDRL research [1].

The treatment of suspicious or confirmed cases must be instituted according to the orientation of the Ministry of Health reference manual [1]. All NBs must be followed-up at outpatient level for the follow-up of titles.

Case report

A mother of a 2-month old female infant, 5.9 kg, sought assistance in an emergency hospital because she observed edema of the right lower limb and irritability to the limb's mobilization in her daughter, believing that the symptoms were related to trauma. The X-ray of the right leg showed a bone lesion in distal fibula. The infant was sent to the Pediatric Oncology outpatient clinic at the Porto Alegre University Hospital for suspicion of neoplasm. During the consultation, lower limb edema with pain in mobilization was observed. She also showed nummular erythematous-squamous lesions in neck and other scar-like lesions in back and left groin (Figure 1). Other aspects of the physical examination without particularities.

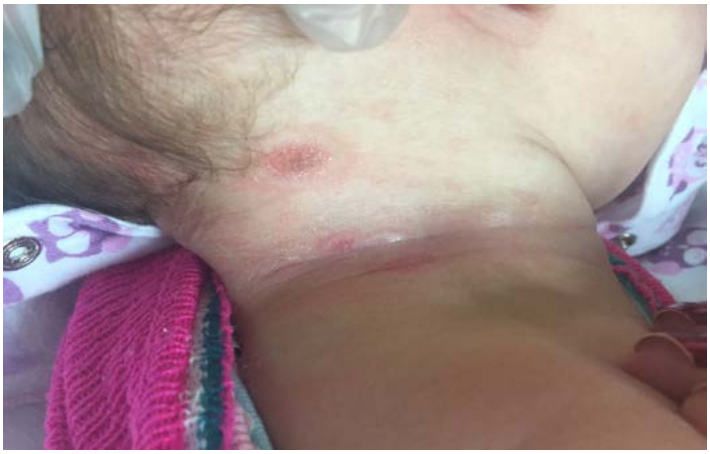

Figure 1: Cutaneous lesions of CS.

Perinatal data: 21-year old mother, primipara, gestational age 38 weeks, prenatal care carried out with a doctor from health insurance plan, 7 appointments, negative serology at 10 and 30 weeks of gestational age. The mother did not collect the requested tests at the end of the third trimester of pregnancy. The tests were not examined nor were new tests requested when she was admitted for childbirth. The infant was born by elective cesarean section on 05/03/2016 at a private hospital in Porto Alegre, Apgar score 9/10, weight 3405g. The infant was released from the hospital 48 hours after being born without complications. Up until 2 months old, she had had only one childcare appointment at a health clinic with a nurse, without alterations. No appointment was carried out with a pediatrician.

During the appointment with the pediatric oncologist, a blood count, coagulation tests, STORCH group serology, and an abdominal ultrasound were requested. Infant was sent for evaluation with an orthopedic staff, who requested a new radiograph of the lower limbs. It showed a single lesion in the right fibula, a rough periosteal reaction in almost the entire fibula associated with the aggressive osteolytic lesion compromising a third of the distal region diaphysis, a well-defined lesion, with cortical bone rupture. It also showed signs of a pathological fracture of the distal fibula (Figure 2). The first hypothesis raised Was eosinophilic granuloma. There were no signs of lesions in other bones.

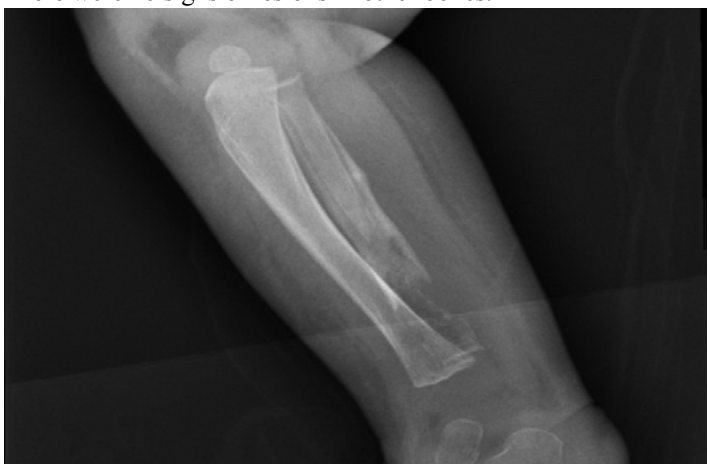

Figure 2: X-Ray right lower limb at admission.

The patient was admitted to the Pediatric Oncology sector on 12/07/2016 to carry out a bone lesion biopsy. Table 1 presents the test results.

Table 1: Laboratory Test Results.

Blood count	Coagulation	Serology	Abdominal ultrasound
HCT= 23.1	PT 12.8	CMV (*) Negative IgG and IgM	Normal
Hb = 7.7	INR 0.91	Toxo Negative IgG and IgM	
Leukocytes = 10,130		VDRL 1:128	
Bands = 2%			
Segs = 31%			
Eosinophils = 3%			
Basophils = 1%			
Monocytes = 7%			
Lymphocytes = 56%			
Platelets = 638,000			

(*) CMV = Cytomegalovirus

The infant then received the CS diagnosis, with bone manifestations, skin lesions and anemia, and was transferred to the pediatrics team for further investigation and ongoing treatment. Lumbar puncture was performed, showing 13 leukocytes (mononuclear prevailing), other aspects normal and negative VDRL.

Patient remained hospitalized for 14 days for antibiotic therapy with Ceftriaxone 100mg/kg/day, due to the lack of Crystalline Penicillin at the hospital. Patient underwent ophthalmological evaluation, which was normal. On 07/15, a plaster cast was put on the right lower limb, which was kept until 08/09/16, when she presented consolidation of the bone lesion in a control X-ray. She was released from the hospital on 07/27/2016 with return for outpatient follow-up. During the follow-up, until 9 months of age, the patient presented a gradual decrease of titles up to 1:8, improvement in anemia and no other clinical complications.

Discussion

CS is responsible for high perinatal morbidity and mortality, with combined rates of miscarriage, fetus and neonatal death reaching 40% [5]. Adequate prenatal care is essential and, in cases of vertical exposure to infection, diagnostic tests and necessary treatment are offered. Even so, the pediatrician must be prepared to identify early clinical manifestations, in order to have a high degree of suspicion of the disease when there are failures in the immediate pre and postnatal identification.

Although most NBs are asymptomatic, mucocutaneous lesions can be present in up to 70% of cases, whether at birth or during the first weeks of life. In this case, lesions began around the third week of life indicating nummular erythematous-squamous lesions in neck, back and groin - the last two already with a scarring aspect when the patient arrived for our appointment. The typical lesions are small, erythematous and maculopapular being most prominent in the back, buttocks and posterior region of the thighs and soles of the feet [6].

Anemia and thrombocytopenia can also be present, often associated with hepatosplenomegaly or splenomegaly [7]. our patient showed anemia (Hb7.7), but with thrombocytosis and without hepatosplenomegaly in the abdominal ultrasound carried out.

Radiological alterations in long bones are common in CS patients, occurring in up to 60-80% of cases. They sometimes can be the only sign of the disease in the NB [6]. they usually appear symmetrically in several long bones. Bone impairment is painful, usually leading to the decrease in limb mobilization (Parrot's pseudoparalysis) [6,8]. Due to the increasing incidence of CS, we highlight the importance of taking it into consideration in the differential diagnosis when we are facing a patient with bone impairment, as in the case of this patient.

As previously explained, the analysis of cerebrospinal fluid (CSF) is essential for the proper handling of CS. For NBs, the CSF is considered as altered with over 25 leukocytes/mm^3, proteins above 150mg/dL, and/or reagent VDRL. For babies more than 28 days of age, as in the case presented, the CSF is considered abnormal when over 5 leukocytes/mm^3, proteins above 40mg/dL and/or reagent VDRL [1]. According to the Ministry of Health protocol, patients with altered CSF must be necessarily treated with intravenous Crystalline Penicillin for 10 days. In the case of lack of Penicillin, these patients must receive Ceftriaxone for 10 to 14 days. In addition to these peculiarities, patients with altered CSF must undergo a new lumbar puncture every 6 months until the normalization of the test.

The Ministry of Health protocol also recommends ophthalmological, neurological and audiological follow-up semiannually for two years, for children with CS diagnosis. Our patient underwent the first ophthalmological evaluation, with a normal result. Her neurological development also appeared adequate for her age.

Considering that Penicillin was not available at our attendance, we opted to treat the patient with Ceftriaxone. It is important to remember, however, that the Ministry of Health protocol emphasizes that up to now, there is no scientific evidence of the efficacy of CS treatment with Ceftriaxone, recommending its use only in the total impossibility of Penicillin treatment. These patients must have a strict clinical follow-up until therapeutic success is proven.

Therefore, we face a serious public health issue marked by the ongoing increase of syphilis cases. In addition to this, the lack of Penicillin contributes to the alarming data regarding CS. Individually we have little to contribute to the solution of this issue immediately. However, in this context we highlight the importance of the health professional in adequate prenatal care, admission for childbirth, and in the childcare appointments. The case presented evidence of a sequence of problems which reinforce the serious consequences of gaps existing in each of these stages in the health care of the pregnant mother and the baby.

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