CASE REPORT

Congenital syphilis: an emerging emergency also in Brazil

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Abstract

Objective: to describe three cases of congenital syphilis.

Case report: we describe the case of three infants who were first diagnosed with congenital syphilis after the neonatal period, when they required emergency care and were admitted to the pediatric intensive care unit. The first patient presented neurosyphilis and nephrotic syndrome; the second presented neurosyphilis; and the third presented hepatitis. We discuss the clinical aspects of these cases as well as other clinical manifestations of congenital syphilis that may lead to emergency situations. We analyze the reasons that may contribute to the failure of diagnosis at birth, and describe some risk factors for gestational syphilis.

Conclusions: in view of the increasing incidence of syphilis in Brazil, and of the possibility that congenital infection is not diagnosed at birth, physicians providing emergency care must keep in mind the maternal risk factors, possible errors in prenatal serology, and the varied clinical presentations of congenital syphilis that might develop during the first months of life.


Introduction

In the past decade we witnessed an increase in the cases of congenital and gestational syphilis in Brazil.1 A similar phenomenon occurred in the late 1980s and early 1990s in the United States and in other developed countries. In those countries, such increase caused significant mobilization, initially in the academic environment, with research to describe and identify the problem,2 and then in public health services, with the implementation of measures aiming at controlling congenital syphilis.3 In developing countries, the incidence of syphilis, in its several forms, continues to increase, reaching the proportion of a true epidemics. Currently, syphilis is considered as a reemerging disease.1

More than 50% of the patients diagnosed with congenital syphilis are asymptomatic at birth. However, the symptoms will appear, in most cases, between the 3rd and 14th weeks of life. Clinical manifestations may vary from very specific or localized signs to general and unspecific signs, making diagnosis difficult. In addition, clinical manifestations may include mild signs and acute presentations, requiring emergency care.4,

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With the objective of drawing attention to the fact that congenital syphilis may go undiagnosed in the neonatal period, causing infants to seek emergency care at a later time, the present work was designed in two stages: a) first, we describe three presentations of congenital syphilis in the post-neonatal period; b) we carry out a brief, however consistent review of the literature on this subject, approaching some clinical and epidemiological aspects of this disease and emphasizing the need for including congenital syphilis in the differential diagnosis of pediatric emergencies.

Case reports

**Case 1**

A ten-week old patient was taken to the emergency room at Hospital São Lucas by a relative, because the mother was ill. The patient had been presenting diarrhea for 2 weeks and bloody feces for 2 days. Upon physical examination, he was dehydrated and malnourished. Weight was 3,100g (weight at birth: 2,600g). He also presented enterorrhagia. We initiated parenteral hydration. Initial examinations: Hemogram: hemoglobin 8g/dl, leukocytes 13,300/mm³ with 24% of band neutrophils. Platelets: 465,000/mm³. Normal liquor.

Since there was a worsening in clinical status, he child was transferred to the pediatric intensive care unit (ICU) and started on antibiotic therapy (ampicillin and gentamicin). Hemoculture was negative. In the following week, clinical and started on antibiotic therapy (ampicillin and gentamicin). We initiated parenteral hydration. Initial examinations: Hemogram: hemoglobin 8g/dl, leukocytes 13,300/mm³ with 24% of band neutrophils. Platelets: 465,000/mm³. Normal liquor.

On the 8th day of hospitalization the mother came to the hospital, because the mother was ill. The patient had been presenting diarrhea for 2 weeks and bloody feces for 2 days. Upon physical examination, he was dehydrated and malnourished. Weight was 3,100g (weight at birth: 2,600g). He also presented enterorrhagia. We initiated parenteral hydration. Initial examinations: Hemogram: hemoglobin 8g/dl, leukocytes 13,300/mm³ with 24% of band neutrophils. Platelets: 465,000/mm³. Normal liquor.

**Case 2**

A 60 day-old patient examined in an emergency service in the greater Porto Alegre metropolitan area. The child was born in a metropolitan area hospital (2,600g), and who was discharged after 28 days, in stable conditions, still malnourished. He was then followed as an outpatient.

**Case 3**

A 40 day-old patient, born from a normal delivery in a metropolitan area hospital (2,600g), and who was discharged with no events on the 2nd day of life. The mother was 31-years old. This was her 3rd child, and the two previous children had been born alive. She did not received any prenatal care, but saw a physician during her pregnancy because of spots on her skin, which were treated as acarnosis. No examinations were required after the appointment or in the hospital where delivery occurred. One month and 5 days after delivery, the baby started presenting fever, moaning, dry cough, and weakness. Two days later, the child was taken to a health care service, where he was given an antithermic. Three days later, presenting a sudden worsening of his condition, he was taken to the local emergency service, and from there he was referred to Hospital São Lucas by a relative, because the mother was ill. The patient had been presenting diarrhea for 2 weeks and bloody feces for 2 days. Upon physical examination, he was dehydrated and malnourished. Weight was 3,100g (weight at birth: 2,600g). He also presented enterorrhagia. We initiated parenteral hydration. Initial examinations: Hemogram: hemoglobin 8g/dl, leukocytes 13,300/mm³ with 24% of band neutrophils. Platelets: 465,000/mm³. Normal liquor.

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Lucas with suspicion of septicemia. Upon hospitalization, he presented important abdominal distension, hepatosplenomegaly, pallor, hypoactivity, and enterorrhagia episodes. Hemogram: hemoglobin 4.6g/dl, leukocytes 22,900 (blasts 4%, metamyelocytes 3%, rods 10%, segmented 32%, lymphocytes 40%, monocytes 11%). Platelets 47,000/mm³. Normal liquor. Normal chest X-ray. Total bilirubin 7.9mg/dl, direct bilirubin 4.7mg/dl, glutamic-oxalacetic (58mg/dl) and glutamic-pyruvic (22mg/dl) transaminase. He was started on broad-spectrum antibiotic therapy and support treatment. On the 5th day we received the following results: serum VDRL 1:64, reagent FTA-Abs and non-reagent liquorrhic VDRL. IV crystalline penicillin 200,000 UI/kg/day was added to the regimen. Hemoculture did not evidence bacterial growth. The child was discharged 15 days later, in regular general state, still presenting hepatomegaly, jaundice, and anemia. He continued being followed as an outpatient.

**Comments and discussion**

The involvement of the central nervous system (CNS) by congenital syphilis is asymptomatic during the neonatal period and sometimes during the first months of life, even in the presence of liquorrhic abnormalities. A symptomatic acute leptomeningitis may appear during the 1st year of life, generally between the 3rd and 6th months. The signs and symptoms may be those of bacterial meningitis; however, liquorrhic findings suggest aseptic meningitis, with moderate degree of pleocytosis, poor proteinorrhaquia, and normal glucose. Liquorrhic VDRL is generally reagent, but its negativity does not rule out CNS involvement. Our first case presented pleocytosis and slight hyperproteinorrhaquia, findings which are compatible with a diagnosis of neurosyphilis, taking into consideration the other characteristics of the case. This form of CNS involvement readily responds to penicillin therapy. Later (generally at the end of the 1st year of life) chronic meningo-vascular syphilis may appear, presenting a protracted course. It may lead to progressive hydrocephaly, cranial nerves paralysia, cerebral vascular lesions, and progressive intellectual deterioration. Rare cases of cerebral infarct resulting from syphilitic endarteritis have been described, presenting as acute hemiplegia, which is generally complicated by convulsive crises.4

Nephrotic syndrome associated with congenital syphilis generally appears between the 2nd and the 3rd months of age. Signs include pre-tibial, escrotal, and palpebral edema, and ascites. Proteinuria and hypoproteinemia characterize the nephrotic syndrome. Nephritic involvement is a consequence of glomerular lesion, which results from damage to immune complexes in the basal membrane. There is interstitial perivascular inflammatory infiltrate, predominantly plasmocytes and lymphocytes. Glomerular epithelial cells are enlarged in number and volume. Eventually there is a dominance of the nphritic component.4

Syphilitic hepatitis may be part of a severe combination of symptoms, as in the septicemic form, or it may appear in isolation. It presents as hepatomegaly, with or without splenomegaly, jaundice with conjugated bilirubin increase, increased hepatic enzymes, and hemorrhagic diathesis. It should be stressed that hepatosplenomegaly and jaundice are the most frequent findings on physical examinations in symptomatic congenital syphilis during the neonatal period.4 A form of fulminant hepatitis with hepatic calcifications has been described, secondary to congenital syphilis.6

Hematological findings such as anemia, low platelet count, and leukocytosis, are rather frequent in congenital syphilis. Anemia is present in almost every case; initially the hemolytic component is more accentuated, while chronic anemia due to medullar hypoplasia appears later on, strengthened by the infant’s physiological anemia.4 Pohl et al.7 described the case of a 5-month-old infant who developed hepatomegaly, intense anemia, and low platelet count. The child also presented maculopapular exanthema. Medullar biopsy revealed marked hematophagocytosis. Serology confirmed the presence of congenital syphilis. Both parents were infected and had not received treatment.

Digestive bleeding occurred in 2 out of the 3 cases reported here, as in a case of death by congenital syphilis described by us previously.8 Probably, low platelet count is involved as a causal factor in most of the cases; hemorrhagic diathesis due to hepatic insufficiency may be involved in some cases. However, there have been reports of syphilitic ileitis due to inflammatory infiltrated in the particular and submucous blade, with mucous ulceration, causing intestinal obstruction and enterorrhagia, as a consequence of congenital syphilis.9

Although none of our patients presented osseous involvement, osseous lesions due to congenital syphilis are one of the most frequent reasons for visits to the emergency rooms. The most common findings are osteochondritis and long bone periostitis. The former takes 5 weeks to appear in radiologic examinations and, the latter, 16 weeks. Thus, it is possible that these lesions are not visible at birth, or, that they are present, but in an asymptomatic form. If the newborn does not receive adequate treatment, osseous lesions may develop into a painful, symptomatic disorder known as Parrot’s pseudoparalysia. In some cases resolution is spontaneous, without symptoms. However, these lesions often lead to pathological fractures, due to demineralization. In emergency trauma care, these fractures may be attributed to abuse; therefore, an accurate diagnosis is essential, in order to avoid erroneous management.10 Benzathine penicillin is not efficient in the treatment of osseous lesions; we have found, in our pediatric practice, cases of Parrot’s pseudoparalysia in infants who had received benzathine penicillin during the neonatal period, which is in agreement with previous reports.4

In 1990, Dorfman et al.5 reported seven cases of congenital syphilis that appeared between the 3rd and the
Congenital syphilis: an emerging emergency... - Lago EG et alii

14th weeks of age, leading to emergency room visits. Four patients presented skin lesions characteristic of congenital syphilis, but three patients had no typical manifestations and presented fever, with no signs and with liquorthic alterations suggesting aseptic meningitis. Most infants presented hepatomegaly and anemia. Along with the article, an editorial emphasized the importance of considering congenital syphilis in the differential diagnosis of infants presenting fever, at least in regions with a high incidence of syphilis.2

Other works published during the past decade report cases of congenital syphilis that were not diagnosed during the neonatal period, later on presenting sudden, seven, and even fulminant events, requiring from the emergency medical team a high degree of suspicion to allow implementation of early treatment. Clinical manifestations included diarrhea, dehydration, fever, respiratory insufficiency, cardio-circulatory arrest, convulsive crises, and several others, as previously described.4,11,12 In 1991, Rosenberg,12 in the United States, classified congenital syphilis as being an "emerging emergency," which inspired the title of the present paper.

Late congenital syphilis, much less frequent, generally presents a less fulminant clinical status, and it is sometimes the cause for appointments in emergency services. Some cases require the differential diagnosis with the acquired infection. In these cases, we should always consider the possibility of sexual abuse.4

The reasons why a case of congenital syphilis is not properly diagnosed and treated, from the neonatal period up to the immediate neonatal period, deserve some considerations.

Current recommendations for the control of congenital syphilis include the performance of VDRL in the first prenatal appointment, with a repeat test at the beginning of the 3rd semester and at birth.1,3 Adequate treatment for pregnant women is the best alternative, since this will prevent fetal infection, or promote cure before delivery. If prenatal treatment is not perfomed, the child must receive treatment as soon as possible during the neonatal period. The diagnosis of congenital syphilis in the newborn cannot be based on clinical data only, since more than half of the infected patients do not present any signs during the neonatal period. Thus, it is necessary to take into account both the serological tests of both the mother and the newborn, the history of maternal treatment. No newborn should be discharged from the maternity ward without a result of maternal VDRL at birth. In addition, a clinical investigation should be performed when indicated, with the appropriate measures depending on the results.1,3,4

In the three cases reported here, the mothers did not receive prenatal care. The absence of prenatal care is considered as the most important risk factor for congenital syphilis in several parts of the world. In one of the cases, there was also some evidence of sexual promiscuity, another risk factor associated with sexually transmitted diseases.4,13 It is important that the risk factors be recognized and taken into account, because they may lead to a diagnostic suspicion in less obvious cases.

In the three cases, the recommendation that no newborn should be discharged from the maternity ward without the confirmation of maternal serology results was not followed. In two cases, a serological test was not even performed, despite the fact that the mothers had not received prenatal care. The verification of VDRL at birth is not only important for those mothers who did not receive prenatal care; it is also important for mother with a previous non-reagent VDRL, since infection or positivity may have occurred later on. Non-treponemal tests become reagent within 4 to 8 weeks after syphilis was acquired. In primary syphilis, non-reactivity of VDRL may be as high as 25%. Thus, an infection acquired at the end of pregnancy may be too subtle to be detected. Again, it is important to stress that frequently primary syphilis does not present clinical signs, mainly in women.4,14

Another factor that may contribute to an unmade syphilis diagnosis is the prozone phenomenon, which occurs when an excess of serum antibodies prevents the formation of the antigen-antibody complex on the slide. Without that, the flocculation reaction can not be visualized. In these cases, serum dilution may avoid a false-negative result. The prozone phenomenon occurs in approximately 2% of primary and secondary syphilis cases.14

Jonna et al.15 described three cases of congenital syphilis that were not diagnosed during the neonatal period. All the mothers presented risk factors, such as absence of prenatal care and use of illegal drugs. Both mothers and babies were sero-negative for syphilis at birth, but the infants became sero-positive when at 2 months old. Those authors reviewed the literature and reported another 73 cases of congenital syphilis diagnosed in the post-neonatal period, when clinical signs are already present making the patient’s prognosis worse. Those authors even suggest that infants from sero-negative mothers who present risk factors be tested for congenital syphilis between the 4th and the 8th weeks of age.

Conclusions
The incidence of congenital syphilis is increasing. In some cases, it may go undiagnosed in the neonatal period. Pediatricians must be prepared to include this disease in the differential diagnosis of several clinical disorders requiring emergency care during the first weeks and months of life. Maternal history is important to corroborate the diagnostic hypothesis, since it may reveal the presence of risk factors that could be associated with the presence of congenital syphilis.
References


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