



Acute Hepatic Insufficiency Disclosing Congenital Syphilis in a Neonate

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Authors' contributions

This work was carried out in collaboration between all authors. Authors AB and MM designed the study, wrote the protocol, and wrote the first draft of the manuscript. Authors MAR, YT, HB and HA managed the literature searches, analyses of the study. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Congenital syphilis is a maternal-fetal infection caused by *Treponema pallidum*. Early signs are characteristic skin lesions, lymphadenopathy, hepatosplenomegaly, failure to thrive, blood-stained nasal discharge, perioral fissures, meningitis, choroiditis, hydrocephalus, seizures, intellectual disability, osteochondritis, and pseudoparalysis (Parrot atrophy of newborn).

We report the case of a new born on day 1 of life hospitalized for respiratory distress. Clinical examination showed signs of respiratory retraction with hypotonia. The assessment carried out revealed: severe liver insufficiency and infected cerebrospinal fluid (CSF) with sterile culture. The

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diagnosis of congenital syphilis in our patient was confirmed by a positive serology in the neonate and her mother. The outcome was fatal.

Keywords: Congenital syphilis; early; neonate; hepatic insufficiency.

1. INTRODUCTION

Congenital syphilis is a multisystem infection caused by *Treponema pallidum* and transmitted to the fetus via the placenta. Overall risk of transplacental infection of the fetus is about 60 to 80%, and likelihood is increased during the 2nd half of the pregnancy. Untreated primary or secondary syphilis in the mother usually is transmitted, but latent or tertiary syphilis usually is not. In neonates, manifestations of syphilis are classified as early congenital (ie, birth through age 2 yr) and late congenital (ie, after age 2 yr). Approximately 66% of infected infants from congenital syphilis are asymptomatic at the time of birth and are identified only by routine prenatal screening [1,2].

Clinical signs appear in approximately two-thirds of affected infants from 3rd to 8th week of life and in most cases by three months of age [1]. Early manifestations are rare.

We are reporting one such case of symptomatic congenital syphilis that presented with liver manifestations soon after birth, a rare finding in literature.

2. PRESENTATION OF CASE

This is a new born on day 1 of life hospitalized for respiratory distress. The mother is 33 years old, treated for primary syphilis discovered at 26 weeks of amenorrhea (WA) and treated by penicilline as per guideline. There is no subsequent serological screening. The husband's serological status is unknown. Pregnancy was followed and aged 36 WA. The delivery was normal, Apgar was 10/10, weight was 1900 g and head circumference 31 cm. Clinical examination immediately after birth showed signs of respiratory retraction with hypotonia, also noted the review has neither rash nor rhinitis. There was no rise of temperature. Anterior fontanel was flat, and there was no asymmetry of movements or cranial nerve palsy.

At hospitalization the patient was treated with ampicillin and gentamicin at a dose of 200 mg/kg/day and 3 mg/kg/day respectively.

Investigations revealed leukocytosis to 26,000 /mL; platelets 39000 /mL; CRP 11.9 mg/L, severe liver failure: Level of prothrombin was 10% and factor V was 25%. Blood cultures was sterile. Cerebrospinal fluid analysis showed low level of sugar (0.06 g/L), high level of protein (3.39 /L) and lymphocytosis 90 cells /mm³. Further, VDRL (Venereal Disease Research Laboratory) test was positive in the baby and the mother with titers 1:960 and 1:128, respectively. *Treponema pallidum* haemagglutination (TPHA) test was positive in the baby and mother. CSF VDRL test was negative. Father's VDRL and TPHA test was negative. Serologies (HIV, HBV, HCV, cytomegalovirus and herpes) were negative. Those tests has realized after consent of the patient party.

On radiological plan, cranial ultrasound showed a choroid plexus hemorrhage. A brain scan revealed a subdural hematoma.

The infant was managed with crystalline penicillin 50,000 units/kg/dose three times a day, oxygen and vitamin K1.

The evolution was marked, 24 hours later, by a generalized icterus with seizures. The control tests showed thrombocytopenia and persistent liver failure despite antibiotics, red cells, plasma transfusions and vaproic acid. At the 11th day of hospitalization the patient dies by hemorrhagic shock.

3. DISCUSSION

Congenital syphilis is acquired by an infant from an infected mother by transplacental transmission of *Treponema pallidum* during pregnancy or possibly at birth from contact with maternal lesions [2]. Intrauterine infection with *Treponema pallidum* can result in still birth, hydrops fetalis, or preterm birth, or be asymptomatic at birth. Early form of congenital syphilis is when the clinical manifestations occur before two years of age and late congenital syphilis is when manifestations occur after two years of age.

General clinical manifestations seen in infants are hepatosplenomegaly, snuffles,

lymphadenopathy, mucocutaneous lesions, pneumonia, edema, rash, haemolytic anemia, or thrombocytopenia at birth or within the first 4–8 weeks of age. Many patients are asymptomatic, and the infection may remain clinically silent throughout their life [1,3].

Early congenital syphilis commonly manifests during the first 3 months of life. Manifestations include characteristic vesiculobullous eruptions or a macular, copper-colored rash on the palms and soles and papular lesions around the nose and mouth and in the diaper area, as well as petechial lesions. Generalized lymphadenopathy and hepatosplenomegaly often occur [4-7].

The infant may fail to thrive and have a characteristic mucopurulent or blood-stained nasal discharge causing snuffles. A few infants develop meningitis, choroiditis, hydrocephalus, or seizures, and others may be intellectually disabled. Within the first 8 months of life, osteochondritis (chondroepiphysitis), especially of the long bones and ribs, may cause pseudoparalysis of the limbs [4,7,8,9].

Diagnosis is usually suspected based on maternal serologic testing, which is routinely done early in pregnancy, and often repeated in the 3rd trimester and at delivery. Neonates of mothers with serologic evidence of syphilis should have a thorough examination, darkfield microscopy of any skin or mucosal lesions, and a quantitative nontreponemal serum test (eg, rapid plasma reagin [RPR], Venereal Disease Research Laboratory [VDRL]); cord blood is not used for serum testing because results are less sensitive and specific [10-12].

Diagnosis is confirmed by microscopic visualization of spirochetes in samples from the neonate or the placenta. Diagnosis based on neonatal serologic testing is complicated by the transplacental transfer of maternal IgG antibodies, which can cause a positive test in the absence of infection. However, a neonatal nontreponemal antibody titer > 4 times the maternal titer would not generally result from passive transfer, and diagnosis is considered confirmed or highly probable.

All seropositive infants and those whose mothers were seropositive should have VDRL or RPR titers every 2 to 3 months until the test is nonreactive or the titer has decreased 4-fold. In uninfected and successfully treated infants, nontreponemal antibody titers are usually

nonreactive by 6 months. Passively acquired treponemal antibodies may be present for longer, perhaps 15 months. [10,12,13].

In confirmed or highly probable cases, 2010 Centers for Disease Control and Prevention (CDC) guidelines recommend aqueous crystalline penicillin G 50,000 units/kg IV q 12 h for the first 7 days of life and q 8 h thereafter for a total of 10 days or procaine penicillin G 50,000 units/kg IM once/day for 10 days. If ≥ 1 day of therapy is missed, the entire course must be repeated [1,4,7,13]. This regimen is also recommended for infants with possible syphilis if the mother fits any of the following criteria:

- Untreated
- Treatment status is unknown
- Treated ≤ 4 wk before delivery
- Inadequately treated (a nonpenicillin regimen)
- Maternal evidence of relapse or reinfection (≥ 4 -fold increase in maternal titer)



Fig. 1. Brain scan meningeal enhancement

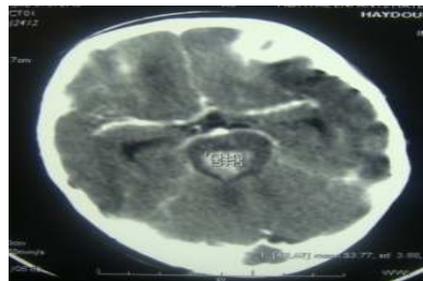


Fig. 2. Abscesses in the brain stem

4. CONCLUSION

Congenital syphilis is a maternal-fetal infection with various presentations. The diagnosis is difficult without a specific context of parental syphilis. It is easily treatable, but can have severe consequences in short and long term

above all the neonates in the absence of appropriate treatment.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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