

Case Report

Congenital Syphilis: Case Report and Review of the Literature

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ABSTRACT

Congenital Syphilis (CS) is rarely reported from this area of world. We report here a case of CS which was diagnosed and treated successfully in Kuwait, describing the clinical implications of this preventable disease and stressing the need for early diagnosis and management to

prevent morbidity and fatality and for prevention of this serious disease at any cost through health care, health education and antenatal screening. Prevention is better than cure.

KEYWORDS : penicillin G, *Treponema pallidum*

INTRODUCTION

Although syphilis was first recognized as a distinct syndrome in Europe in the 15th century, it still poses a serious health problem in many countries in spite of the wide-spread use of penicillin to treat syphilis since the early 1950's^[1-3]. A case of congenital syphilis is defined as an infant born to a mother who had inadequately treated or untreated syphilis at delivery or any infant with a reactive treponemal test and any symptoms or signs of syphilis on physical examination^[4,5]. The etiologic agent of syphilis is a spirochete named *Treponema pallidum*. The infection is commonly transmitted horizontally by sexual contact and less so vertically through hematogenous spread across the placenta. The incubation period for acquired primary syphilis ranges from 10 to 90 days^[6,7].

We are reporting a case of congenital syphilis, describing the clinical implications of this disease and emphasizing that global antenatal screening is still mandatory to prevent this serious, yet largely preventable, disease in high-risk mothers.

CASE REPORT

The mother presented a male non-Kuwaiti, born in August 1998, to our unit at the age of 60 days with poor weight gain and mild pallor. He is a product of full-term normal vaginal delivery from non-consanguineous GCC parents living outside Kuwait. His birth weight was 2.3 kg. He had a history of neonatal jaundice on the first day of life, which was treated conservatively. Both parents had a history of treatment for syphilis. The father has also a history of previously treated pulmonary TB. He was not compliant to the treatment of syphilis. Mother was diagnosed to have syphilis late in

pregnancy when she developed lesions on the genital area. She was started on intramuscular penicillin injections 10 days prior to delivery and then maintained on oral penicillin. Our patient has 9 sisters and 4 brothers, all are said to be alive and well. He was breast fed until the age of 15 months and was fully vaccinated. Developmental assessment was normal except for mild speech and walking delay.

Physical examination at presentation revealed weight, height, and head circumference at the 5th percentile, mild pallor, a small umbilical hernia and abdominal distension with liver being felt firm 5 cm and spleen 4 cm below their respective costal margins. The skeletal system revealed a left clubfoot. He had no dysmorphic features or skin rashes. Systemic examination was otherwise unremarkable.

Investigations at two months revealed leucocytosis of $17.2 \times 10^9/L$, polymorphs 20%, lymphocytes 73%, monocytes 3%, eosinophils 4%. Hb 97 g/l with normochromic normocytic red blood cell (RBC) picture, reticulocytes 8%, Platelets $136 \times 10^9/L$, ESR 30 mm/hr, quantitative CRP 52.9 mg/l (RR: <8.0), Calcium was 2.46 mmol/l, Phosphorus 1.00 mmol/l, Total Bilirubin 52 umol/l (RR: 3-20) and Albumin 28 g/l (RR: 37-47). His Alkaline Phosphatase was elevated to a level of 1254 U/L (RR: <250), and ALT 209 U/L (RR: 30-65), (Fig. 1). Immunoglobulin electrophoresis showed initially high levels as follows: IgG 9.23 g/l (RR: 3.0-8.0), IgA 1.42 g/l (RR: 0.01-0.40), and IgM 0.9 g/l (RR: 0.19-0.76). These levels dropped to normal values after treatment. Blood glucose, urea, creatinine, sodium, and potassium continued to be normal with negative Coombs test.

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Rapid Plasma Reagin test (RPR) remained always positive and Treponema Palli Hemagglutinin tests (TPHA) showed initially rising titers (Fig. 2). Blood culture was sterile and blood for HIV and hepatitis screen was negative. Cerebrospinal fluid (CSF) sample was bloody and thus unfit for examination for biochemistry and serology, but routine culture and bacterial antigen screen were negative. The mother refused further attempts to repeat the CSF examination. Skeletal survey revealed no radiological evidence of syphilis apart from clubfoot. Ultrasound examination of the abdomen and pelvis confirmed homogenous hepatosplenomegaly with normal appearance of other organs. Head ultrasound was normal, as was the hip ultrasound. The mother's blood tests were positive for RPR with high TPHA titer but negative for hepatitis screen and HIV. The father was not available for investigations.

In our unit, the patient was started on a three-week course of intravenous aqueous crystalline penicillin G 50,000 U/kg (at 8-hour intervals). He tolerated treatment well without any complications. On follow up visits, the size of liver and spleen regressed to normal and the liver enzymes were back to the normal range by seven months of age. TPHA titers dropped to the lowest level (1/80) by the age of one year.

At the age of 1 year 3 months, he presented with poor feeding, failure to gain weight along with mild hepatosplenomegaly and a four-fold rise in TPHA titer (1/1280). The mother's blood test also showed a significant rise in the serology titer. The mother again refused a CSF analysis in the child. Father was said to be still non-compliant on treatment. The child was given another three-week course of I.V. crystalline penicillin with good response. The mother was seen by her physician who re-started her on penicillin therapy. The child's clubfoot was successfully treated by surgery by an orthopedic surgeon. He was doing well when last seen on follow up in our clinic at the age of 2 years and 10 months, with falling VDRL and TPHA titers and with no evidence of further relapse (treatment failure) or re-infection.

DISCUSSION

In the Middle East, several reports have been published about non-venereal syphilis (Bejel), a contagious, non-sexually transmitted treponematoses of primitive communities which, if not treated, can cause deformities in its late stage. This form indeed had been reported to be endemic among semi-nomadic Bedouins in some parts of Saudi Arabia. In contrast, venereal syphilis is uncommon, and congenital syphilis (CS) is even more rare in this area and is found almost exclusively in urban populations^[8-11]. Although syphilis occurs

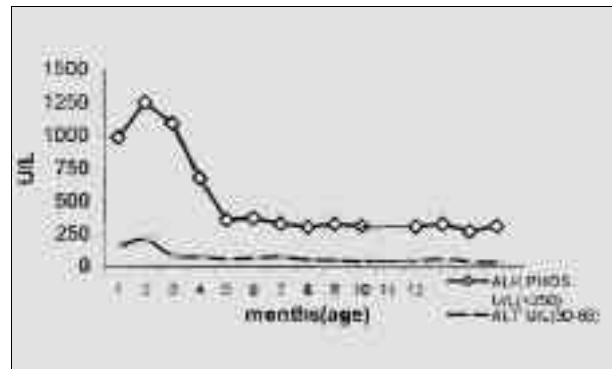


Fig. 1: Liver enzyme levels for the patient

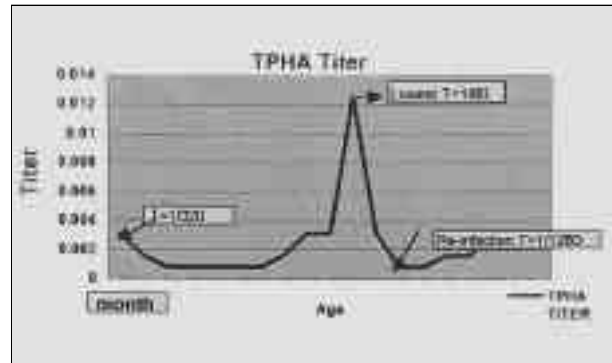


Fig. 2: TPHA titer values for the patient

infrequently among Canadian and American women, global antenatal screening is still warranted. The reason is that congenitally acquired syphilis is serious, yet largely preventable. At least half of infected live-born infants have no signs of congenital syphilis at birth. Many of them were often not reported even though they usually receive presumptive treatment. There were several reasons for not identifying these infants at birth such as: (i) being asymptomatic at birth; (ii) confusion about interpretation of test titers, especially when mothers' titers were positive and infants' were negative; (iii) difficulty to document the adequacy of treatment of mothers who were positive; (iv) newborns usually discharged within 1 day of birth before obtaining RPR titers^[12,13]. It is estimated that of all pregnant women with untreated syphilis, only 20% will have a term delivery of a normal child. The disease in affected fetuses may be complicated by stillbirth (30%), neonatal death (10%) and mental handicap (40%)^[7]. Our patient was not identified at birth as he was born in a private hospital with no information given to the hospital about mother's condition. In addition, he was asymptomatic at birth, and was discharged on the second day of life before doing RPR titer.

CS usually results from transplacental transfer of the spirochetes to the developing fetus. Although extremely rare, syphilis can also be acquired by contact with a chancre at birth^[14]. The majority of infants do not have skin lesions or

snuffles (a profuse nasal drainage that can be bloody). In addition, the placenta and amniotic fluid is often not available for testing. In these cases, the diagnosis may be difficult and will depend on a combination of physical, radiographic and serologic examinations.

The clinical spectrum of early CS is remarkably variable, ranging from asymptomatic infection to fulminant disease.

Laboratory diagnosis of syphilis can be established by identifying the pathogen. Dark-field examination of clinical specimens taken from a moist genital lesion or a regional lymph node may yield *T pallidum* when patients are infected with primary or secondary syphilis. Screening is accomplished by non-treponemal tests such as the Venereal Disease Research Laboratory (VDRL) or the Rapid Plasma Reagin (RPR) tests. When a positive test is obtained with a non-treponemal test, a specific test for anti-treponemal antibodies should be performed to confirm the diagnosis. These tests are the Microhemagglutination Assay for Antibodies to *Treponema pallidum* (MAH-TP) test, which is equivalent to the TPHA test done for our patient; or the Fluorescent Treponemal Antibody-Absorption (FTA-Abs) test.

Serofast patients are often identified by a low titer non-treponemal result and a positive treponemal-specific test despite appropriate therapy. Re-infection in the serofast patient is diagnosed if a four-fold rise in titer occurs. In this situation, the same type of non-treponemal test should be used to make the diagnosis because of the differences in measured titers between RPR and VDRL^[15,16].

The findings in cerebrospinal fluid (CSF) of a positive VDRL or an increase in leukocytes or protein are the hallmarks of neurosyphilis in adults, but the interpretation of results on CSF specimens from the newborn is more difficult because these results are neither sensitive nor specific for neurosyphilis.

A retrospective study was designed in 1996 in Washington, D.C. to evaluate the usefulness of lumbar puncture (LP) in the initial evaluation of symptom-free infants for congenital syphilis. The study concluded that the role of routine LP in the initial evaluation of these infants should be reconsidered because of the low yield of reactive CSF VDRL and the similar CSF leukocyte and protein values in the syphilis group and the control infants^[17]. In spite of this, evaluation of CSF should still be performed to permit abnormalities to be monitored and also because positive CSF may provide the only evidence of congenital syphilis in asymptomatic infants born to treated mothers^[18].

Prenatal diagnosis of fetal syphilis is possible by the use of ultrasonography. Sonographic findings may include hydrops fetalis, hepatosplenomegaly,

placentomegaly, and dilated small bowel. Amniotic fluid examination can be done by either Rabbit infectivity testing (RIT) to confirm presence of *T pallidum* or by the more specific PCR technique^[19-21].

Although penicillin was first used to treat congenital syphilis in the 1940s, many studies have been designed to determine the optimal treatment for this disease. In 1989, both the American Academy of Pediatrics and the Centers for Disease Control (CDC) recommend treating all infants born to women with untreated syphilis with parenteral penicillin regardless of clinical examination or laboratory findings^[22,23]. In 1998, the CDC recommended that treatment is required in the following situations: a confirmed or presumptive diagnoses of CS, unknown or undocumented maternal therapy, maternal treatment within four weeks of delivery, insufficient fall in maternal titer in response to therapy or delivery before a four-fold fall in titer, maternal treatment with drugs other than penicillin, or in situations in which infant follow-up may be inadequate^[24]. CDC recommends giving crystalline penicillin for neonates with CS in a dose of 50000 units/kg/dose intravenously every 8 to 12 hours for 10 to 14 days. This dose is comparable to that normally administered to an infant with bacterial meningitis.

Our patient received a fully extended course of this treatment for three weeks, after which he showed good clinical response. There was a steady fall in the TPHA titers reaching a minimum of 1/80 at one year of age. His four fold rise in TPHA titer (1/1280) at 15 months of age was simultaneous with rising titer in the mother suggesting re-infection. He was still breast-feeding at the time of the rise in the titer. However, it was not clear if the re-infection in the baby was through breast feeding or close contact with the mother whose husband was non-compliant to therapy as well as personal habits. CS is a preventable disease through adherence to the practice of Islam and through health care and health education. Indeed, prevention is better than cure.

CONCLUSION

The diagnosis of congenital syphilis remains complicated and its reporting remains controversial. Because of the high morbidity associated with congenital syphilis, screening of all pregnant women has been shown to be cost effective even in populations with low prevalence. In high-risk populations, serological screening for syphilis at the first prenatal visit, in the third trimester, and at delivery are important and recommended. An additional screening of infants born to high-risk mothers may be appropriate at 4 to 8 weeks of age.

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