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CONGENITAL SYPHILIS IN AN IMMUNOCOMPROMISED NEONATE:CASE REPORT

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SUMMARY

Syphilis is a notifiable and preventable disease, congenital syphilis more so. Consequently, attention has been recently focused on prenatal diagnosis of foetal syphilis by the use of ultrasonography apart from the conventional serologic screening. Congenital syphilis has not been reported from the Kingdom of Lesotho. We report the case of a 3.0kg male neonate with florid joint and bone lesions of congenital syphilis associated with HIV infection seen at the Queen Elizabeth II Hospital, Maseru, Kingdom of Lesotho. Co-existing HIV infection influences the clinical manifestation of syphilis, the progression of neurosyphilis and the response to standard therapy. The baby had the recommended standard treatment with good response and he was followed-up for a period of twelve months with serologic screening and radiographic evaluation.

INTRODUCTION

Although venereal syphilis was first recognised in Europe in the late 15th century(1) and a global campaign for its control with the wide-spread use of penicillin was initiated in the early 1950s, it still remains a serious health problem in many countries(1-4). Syphilis is caused by infection with a spirochete named Treponema pallidum. Humans are the natural host of T. pallidum and also serve as vector. Sexual contact provides the usual means of transmission(5-7). The incubation period for acquired primary syphilis ranges from 10-90 days(1, 5-7). Treponema pallidum does not survive well outside the host; it is easily destroyed by drying and soap and water(1,7).

Congenital syphilis is defined as an infant born to a mother who had untreated or inadequately treated syphilis at delivery or any infant with a reactive treponemal test and any symptoms or signs of syphilis on physical examination(7-9). Babies are usually infected in-utero by transplacental passage of T. pallidum from an infected mother, but infection may also occur by contact with an infectious genital lesion during passage through the birth canal(5-7). It is still unclear what factors determine which mother, particularly those in latent stage of the infection, will pass the disease to their foetuses or why some infants who are infected in-utero are born asymptomatic, but develop overt disease in the first weeks or months of life(7).

The prevalence rate of HIV infection in the Kingdom of Lesotho is about 28%, (10) and syphilis is known to be common in HIV infected adults(7). Consequently, babies of HIV infected mothers are at high risk of contracting both HIV and congenital syphilis; thus our interest in reporting this case.

CASE REPORT

The two-week-old male Mosotho baby had been born at term to a 27-year-old para three full-time housewife. Delivery was reportedly normal by the vaginal route at one of the district government hospitals. The stated Apgar scores were eight and ten at one and five minutes respectively. The birth weight was 3.0kgs. The mother booked late at 30 weeks for antenatal care. The details of her ante-natal care were not available and could not be traced. She volunteered a history during pregnancy of copious yellowish, foul smelling discharge per vaginum and genital ulcers which were resistant to some oral antibiotic treatment given. Her husband, a self-employed building contractor had apparently not been well, suffering from urethral discharge and intermittently suppurative bilateral inguinal Lymphadenitis for about one year.

The baby had been born with bilateral swollen lower limbs worse at the knee joints and had been unable to move both lower limbs since birth. He also developed soon after birth, copious yellowish discharge from both eyes as well as purulent discharge from the umbilicus. The progressively worsening symptoms of the baby necessitated referral to the Queen Elizabeth II Hospital. There was no record of treatment from the referring hospital.

Clinical examination revealed a very ill looking neonate, pink in room temperature and not jaundiced. His body weight was 3.6kgs and temperature, 36.6°C. There was purulent discharge from both eyes and the umbilicus. The respiratory rate was 64/minute with good air entry into both lungs and vesicular breath

sounds only. The heart rate, 148/minute was regular with normal first and second heart sounds. No murmurs were heard. There were no palpable intra-abdominal organomegaly. The bowel sounds were present and normo-active. Both lower limbs were grossly swollen, held in abduction and quite tender. The occipito-frontal circumference was 36.0cm, anterior fontanelle was at and pulsatile, the neck was supple but the reflexes were depressed in the upper limbs.

The results of relevant investigations revealed a normal white blood cell count of 9.5 x 10^9/L, haemoglobin of 10.4g/dl with normochronic, normocytic red blood cell picture. Blood for Venereal Disease Research Laboratory (VDRL) was reactive at 1:128, the Treponema Pallidum Haemagglutination Assay (TPHA) was positive at 1:640. The Human Immunodeficiency Virus (HIV) screening (tested with Abbott third generation, Enzyme Linked Immunosorbent Assay ELIZA) was positive. Immunoglobulin electrophoresis was not done. Swab of both eyes and umbilical discharge grew non-haemolytic streptococcus; sensitive to penicillin, tetracycline, ampicillin, gentamycin and chloramphenicol. The liver function, cerebrospinal fluid, blood culture, urine culture and abdominal ultrasound studies were normal. Radiograph of both femora, tibia and fibulae (Figure 1) showed diffuse osteoperiostitis.

Figure 1

Photograph of antero-posterior X-ray showing severe periosteal reaction around the epiphyseal plate; note the bilateral hyperostosis especially around the proximal femur and tibia

The patient showed remarkable improvement to treatment with the standard dose of aqueous crystalline penicillin G. (150,000 units/kg/day) intravenously, in three divided doses; eight hourly for two weeks. The discharge from both eyes and the umbilicus ceased by the fifth day of antibiotic treatment, the lower limb swellings subsided and the baby was able to move the limbs actively. The baby was discharged after three weeks for subsequent out patient follow-up. There was a progressive drop in the VDRL level to 1:64,1:32, and 1:4 at the age of three months, six months and one year respectively. A repeat radiological assessment of the femora, tibia and fibulae at the age of six months did not show any appreciable change in the bone lesion. Both parents whose blood tests were VDRL reactive and positive for TPHA and HIV were referred to the Special Treatment Clinic (STC) for appropriate management of the venereal disease.

DISCUSSION

The Kingdom of Lesotho enjoys a temperate climate with four distinct seasons; summer, autumn, winter and spring and it is almost entirely free of tropical diseases that are major problems in most of sub-Saharan African countries(11,12). The endemic trepanomatoses-pinta, yaws and endemic syphilis (bejel) are unknown to occur in Lesotho. However, venereal syphilis occurs throughout the world but it is most frequent in large urban areas(1,7)

Congenital syphilis (CS) usually results from transplacental transfer of the treponeme to the developing foetus. Rarely, syphilis can also be acquired by contact with a chancre at birth(5-7,13). It is unclear what factors determine which mother, particularly those in latent stage of the infection, will pass the disease to their foetuses, or why some infants who are infected in-utero are born asymptomatic, but develop overt disease in the first weeks or months of life(7). Generally, untreated syphilis can be transmitted to the foetus at any stage of the disease. The rate of transmission is about 100% during the second stage and slowly decreases with increasing duration of the disease(7). The majority of infants do not have snuffles (a profuse nasal discharge that may be bloody) or skin lesions. In addition, the placenta and amniotic fluid are 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(4). Congenital syphilis has traditionally been divided somewhat arbitrarily into early and late stages. Clinical manifestations appearing within the first two years of life are designated early and those occurring after this time are called late, though there can be considerable overlap(l,7). The clinical spectrum of early CS is remarkably variable, ranging from asymptomatic infection to fulminant disease(4). Our patient with florid joint and bone lesions obviously had an early manifestation of the disease.

A laboratory diagnosis of syphilis can be established by identifying the treponeme (T. pallidum) under dark-field microscope in a clinical specimen taken from moist genital lesion or regional Iymph node in patients infected with primary or secondary syphilis. Screening is accomplished by non-treponemal tests such as the Venereal Disease Research Laboratory (VDRL),which

was reactive in our patient, or the Rapid Plasma Reagin (RPR). A reactive non-treponemal test should be followed by a specific test for anti-treponemal antibodies to confirm the diagnosis. The anti-treponemal antibody tests are Treponemal Pallidum Haemagglutination Assay (TPHA), which was positive in our patient, or Micro Haemagglutination Assay to Treponemal Pallidum (MAHTP), or the Fluorescent Treponemal Antibody-Absorption (FTA-ABS).

A cerebrospinal fluid (CSF) study with reactive VDRL, leucocytosis or elevated protein are pathognomonic of neurosyphilis in adults, but the interpretation of results of analysis of CSF obtained from newborns is more difficult because they are neither sensitive nor specific for neurosyphilis(4,7). Furthermore, the results of a retrospective study in 1996 (14), conducted in Washington D. C. to evaluate the usefulness of lumbar puncture (LP) in the initial evaluation of symptom-free infants for congenital syphilis, revealed a low yield of reactive CSF VDRL. Similar CSF leucocyte and protein values were found in both the syphilis group and the control infants. It is therefore not surprising that the CSF study in our patient was normal. However, 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(7,l5).

In recent times, prenatal diagnosis of foetal syphilis is known to be possible by the use of ultrasonography. The sonographic findings may include hydrops foetalis, hepatosplenomegaly, placentomegaly and dilated small bowel. Furthermore, amniotic fluid examination can be done by either Rabbit Infectivity Testing (RIT) to confirm the presence of T pallidum or by the more specific Polymerase Chain Reaction (PCR) technique(l6-18). These diagnostic facilities were not available in our health institution. However, where available, if judiciously utilised, they may further enhance the prenatal diagnosis of congenital syphilis and the initiation of early and appropriate standard treatment. Thus minimising foetal, and perinatal morbidity and mortality. In spite of the fact that penicillin was first used to treat congenital syphilis in the 1940s, many studies have been designed at determining the optimal treatment for this condition (4). The American Academy of Paediatrics and the Centres for Disease Control (CDC), recommended administration of parenteral penicillin to all infants born to women with untreated syphilis regardless of clinical examination or laboratory findings(19,20). The CDC further recommended that treatment is required in the following defined circumstances viz: a confirmed or presumptive diagnosis of CS; unknown or undocumented 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(2l). The CDC recommends giving crystalline penicillin to neonates with CS in a dose of 50,000 units/Kg/ dose every 8 to 12 hours (or 150,000 units/kg/day in three divided doses) for 10 to 14 days. This dose is comparable to that normally administered to an infant with bacterial meningitis(4). Although treatment may cure the infection, the prognosis in treated cases depends on the degree of damage that has already occurred. In general, the earlier treatment is initiated, the more likely it is that a satisfactory response will be obtained(7). However, treatment in-utero in the presence of marked damage to the foetus may neither prevent abortion, stillbirths, neonatal death nor reverse stigmata that have already appeared. Furthermore, the relationship between adequate treatment of early CS and the long-term outcome is as yet inadequately documented(7). Our patient with confirmed CS born to a mother with unknown therapy and insufficient fall in maternal titer in response to therapy received the recommended standard treatment for two weeks, and showed good clinical response. Although there was residual bony lesion at the age of six months, there was a steady fall in the VDRL titers to a minimum of 1:4 at one year of age. It is not unlikely that the immune status of our patient influenced the early and florid manifestation as well as the residual bony lesion.

In conclusion, congenital syphilis is an eminently preventable disease. In order to minimise the likelihood of its occurrence, every woman who becomes pregnant should have a serologic test for syphilis during the first trimester. A repeat test towards the end of gestation or at the time of delivery should be done for women in high-risk groups. The cord blood should also be routinely tested. Cases detected in-utero or after birth should have supervised standard treatment and follow-up evaluation. Case registers regarding CS should be well maintained.

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