Congenital syphilis presenting as dactylitis

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Summary:

An 11 months old male child presented to the STD clinic, Colombo with dactylitis of middle phalanx of middle and ring fingers of both hands. Radiological and serological tests confirmed the diagnosis of congenital syphilis. Baby received inward treatment with a ten day course of penicillin. Both parents were subsequently diagnosed to have early latent syphilis and were treated adequately with penicillin. Parents and baby had no evidence of other STIs.

Introduction

In Sri Lanka the prevalence of infectious syphilis is observed to be decreasing although incidence cases of congenital syphilis have been observed during the last ten years. High-risk behaviors among the sexually active age group continue in the country making women vulnerable to sexually transmitted infections (STIs) including human immunodeficiency virus (HIV). Although antenatal screening for syphilis is compulsory coverage across the country is poor. Venereal Disease Research Laboratory test (VDRL) is the commonly used screening test and if positive has to be confirmed by a specific treponemal test. Treatment early in gestation before significant foetal development has taken place will minimize adverse pregnancy outcomes such as abortion, stillbirths, and congenital syphilis ⁽¹⁾.

Untreated syphilis in the antenatal mother may result in congenital syphilis. Most reports conclude that *Treponema Pallidum* cross the placenta during the 2nd trimester resulting in several adverse outcomes such as abortion, stillbirth or even congenital syphilis. Congenital syphilis is divided into two stages. Early congenital syphilis is characterized by the appearance of signs and symptoms before the age of two years. Some infants may be born with signs and symptoms of congenital syphilis. In some, clinical signs begin to appear in 3rd- 8th week of life. When the onset of infection is in the first few weeks of life usually the prognosis is poor. The most characteristic features are prematurity and low birth weight, skin eruptions, and hepatomegaly, with or without splenomegaly. Other features reported are anaemia, poor

feeding, elevated liver enzymes, pneumonia, jaundice, generalized lymphadenopathy and failure to thrive. Late congenital syphilis is when the diagnosis is made after 2 years of age usually around puberty ^(1, 2, &3).

The diagnosis is by identification of treponemes using dark field microscopy (DFM) or direct fluorescent antibody stain (DFA), staining of treponemes in histology specimens, and by serological tests ⁽⁷⁾.

The cost effectiveness of screening tests for syphilis will depend on the prevalence in the population and in the risk groups. Non-treponemal tests - VDRL or Rapid Plasma Reagin test (RPR) is useful for screening infectious syphilis. However, it will fail to diagnose early primary and late latent or late syphilis. In addition biological false positives are reported with non-treponemal tests and false negative results in the presence of the prozone phenomenon in secondary syphilis.

Antenatal screening is cost effective even in low prevalence countries as screening enables treatment in pregnant women and prevent adverse pregnancy outcome ⁽⁷⁾. Penicillin is the drug of choice for pregnant mothers and for babies infected with syphilis ^(1, 2, 3, 7 &8).

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An 11 months old male baby, first child in the family had a negligible epidemiological-clinical history.

Initially this child was presented to a general hospital in the western province with a two-week history of painless swelling of the middle & ring fingers of left hand (Fig1). It was not preceded by or followed by fever generalized or localized rash of the body, running nose, failure to thrive, loss of weight & recurrent respiratory tract infections (RTIs). But it was followed by gradual involvement of the right hand. On examination there were no signs and symptoms of inflammation of the joints of fingers. Swelling of soft tissue of the middle phalanx of both fingers of both hands were present. Mild pallor and hepato splenomegaly were noted.

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Figure 1. Soft tissue swelling of the middle & ring finger of the Left hand.

Figure 2. Soft tissue swelling seen in 'X' ray of Left hand.

This child has had an uneventful full term vaginal delivery and his birth weight was 2.9 kilograms and was immunized according to the schedule. Investigations showed;

weight	7.7kg.
WBC	12700/ml, N-26%, L-72%, E-2%,
Platelets	453X10 ⁹ /l.
Hb	10.0g/dl
blood picture	hypochromic, polychromic cells with no blast cells
ESR	30mm at 1 st hour

X' ray L/ hand (Fig 2) showed features of dactylitis. Mantoux test was positive at 10mm, with BCG scar. Tuberculosis was excluded on further investigation.

The child was transferred to the Rheumatology ward, of the children's hospital, Colombo for specialized care. Repeat investigations showed;

WBC	10800 N-32%, L-61%, E-7%,
Platelet	48500
Hb	10.4g/dl
blood picture	Features of iron deficiency anemia with hypochromic microcytic red cells with anisocytosis.
ESR	55mm at 1 st hour

Periostitis of humerus and tibia confirmed the diagnosis of syphilis (Fig 3 & 4).

Figure 3. Periosteal reaction in 'X' ray Of long bones (tibiae)

Figure 4. Periosteal reaction in 'X' ray of humerus.

The non-treponemal Venereal Disease Research Laboratory (VDRL) test done at the NSACP was reactive in 512 dilution (R-512) and the treponema specific Treponema Pallidum Partition Agglutination test (TPPA) was positive 2+, which is the maximum level of reactivity (TPPA-2+). The Fluorescent Treponemal Antibody absorption test for IgG (FTA abs IgG) was-4+, and for

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IgM (FTA abs IgM) was 1+ indicating infection of the baby with *Treponema pallidum*. Eye screening, 2D echo, US scan of abdomen & pelvis were normal. There were no other sexually transmitted infections (STIs) identified on screening the baby.

At lumbar puncture (LP) the opening pressure was normal with clear fluid. Analysis of Cerebrospinal Fluid (CSF) showed, clear appearance, proteins-18mg%, sugar-2.4 (NL 3.8-5.6), [random blood sugar (RBS)-5.2 (NL 3.8-5.6)], Microscopy showed no cells and, no organisms on Gram stain, & CSF culture for bacteria, mycobacterium and fungi was negative. CSF- VDRL was non reactive- (NR), FTA abs IgG-2+, & FTA abs IGM-negative.

Child was treated with crystalline penicillin 50,000U/kg (375,000 units) intravenously (IV) 6hourly for 10 days⁽⁸⁾.

The mother had no past history of adverse pregnancy outcomes such as abortion, stillbirths. She gives no history of blood transfusions. Mother had attended both local & hospital antenatal clinics regularly from 13 weeks of gestation (POA). VDRL test carried out at 15 weeks of POA in a private clinic revealed non-reactive results (VDRL-NR).

Both parents were healthy and did not have illness suggestive of primary or secondary syphilis prior to this incident and there were no histories of other STIs in the past.

Mother is an ex- garment factory worker while the father is presently working as a supervisor in a garment factory.

Both parents denied premarital & extramarital sexual exposures. On investigation the mother's serology revealed evidence of treponemal infection. Her VDRL test was reactive in 64 dilutions (R-64) and the father was reactive in 32 dilutions (R- 32). Both had a high positive reaction in the TPPA test (TPPA- 2+) indicating early treponemal infection. Both were treated for early latent syphilis with Benzathine penicillin 2.4 MU IM in a single dose.

They are being followed up at the clinics. Follow up VDRL test of the baby shows a gradual decline in the titer. The parents follow up serology also shows a decline indicating response to treatment.

Discussion

Syphilis is a chronic, systemic infection⁽⁴⁾. Lesions are mainly due to vascular changes leading to endarteritis and periarteritis^(4, & 5).

Vertical transmission is an important feature in syphilis. The risk of vertical transmission is related to the stage of maternal infection. Of infants born to mothers with primary or secondary syphilis, up to 50% will be premature, stillborn, or die in the neonatal period, with others developing signs of congenital disease. Of infants born to women with syphilis in the early latent phase, approximately 35% will be premature, stillborn, or die in the first year of life, another 40% will develop signs of congenital disease. In late latent syphilis, 10% of infants develop signs of congenital disease and up to 10% may be stillborn⁽⁵⁾. This indicates the importance of screening pregnant mothers in pregnancy.

Effective screening of pregnant women for syphilis can be done within a well functioning good quality antenatal care service. Congenital syphilis can be prevented by testing women for syphilis early in pregnancy, treating those who are seropositive, and preventing reinfection. Although most countries have a policy of antenatal syphilis screening, the implementation of programs has made difficult for various reasons. Lack of awareness of service providers, lack of training and logistical support for service providers, difficulty in performing classical tests, non suitable tests, non availability of test results on time, are some of the reasons for the failure of the program⁽⁶⁾.

Screening for syphilis is performed with a non-treponemal test (e.g., rapid plasma reagin [RPR] or VDRL test) in the first trimester. Ideally those at high risk should be retested at 28 weeks and again near the time of delivery⁽⁶⁾. However identifying high risk women is difficult in the community. The non treponemal test may be negative during incubation or in early primary syphilis, so that even with appropriate screening some cases can be missed. This is a recognized reason for failure to treat during pregnancy⁽⁵⁾.

At the time of presentation, in addition to the clinical symptoms this baby's VDRL test was highly reactive with, both IgG and IgM specific treponemal tests being positive. The IgG positivity may reflect the passively transmitted maternal antibodies through the placenta; the IgM positivity indicated active infection in the child⁽⁹⁾.

The diagnosis of congenital syphilis was confirmed when the FTA-abs IgM test on the baby's blood was positive⁽⁹⁾.

In this case mother's non reactive VDRL test during the first trimester of pregnancy can be explained in different ways; (a) prozone phenomenon, (b) mother acquiring the infection later in her pregnancy, (c) mother is in the

incubating period, and (d) laboratory errors. This justifies the need of a repeat antenatal VDRL test in second or third trimester. Financial and logistical constraints and lack of awareness of the health care providers, hinder performing a repeat VDRL test in antenatal clinics ⁽⁶⁾.

By far the greatest value of this case report is its missed opportunity to diagnose early latent syphilis in the mother and providing appropriate treatment on time to prevent congenital syphilis. Therefore this case report may prevent such occurrences in the future.

Congenital syphilis elimination strategy in our country should include improving the existing policy with widening the availability of screening tests, improving knowledge of the health care workers and parents on screening, acquisition, and treatment of congenital syphilis ⁽⁶⁾.

Congenital syphilis in our times is preventable, and all efforts and control measures should be taken to prevent such a situation in the future.

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