

EARLY CONGENITAL SYPHILIS: EXPERIENCE WITH 13 CONSECUTIVE CASES SEEN AT THE UNIVERSITY HOSPITAL, KUALA LUMPUR

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ABSTRACT

While it is not difficult to recognise the classical clinical features of congenital syphilis in most cases, some of them may present with unusual manifestations which can defy early diagnosis. We report our experience with 13 cases of early congenital syphilis over a period of 10 years from 1980 to 1989. Twelve of the thirteen patients were less than 3 months at presentation. There were two infants born prematurely and six of the babies were born with a low birthweight (less than 2.5 kg). All but four patients survived following treatment.

Skin lesions either in the form of typical vesiculobullous eruption over the palms and soles or a maculopapular skin rash over the body were the most common presentation and was seen in 10 patients. Splenomegaly with or without hepatomegaly was the most consistent physical sign. Radiological changes in the form of periostitis and/or metaphysitis were seen in all cases where an X-ray of the long bones was performed. An elevated serum immunoglobulin M, though non-specific for the disease, was found to be a useful screening test for recent infection.

Keywords : Early congenital syphilis, periostitis, metaphysitis, immunoglobulin M, splenomegaly

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INTRODUCTION

Over the last decade or so the medical scene has been dominated by research into finding a cure and a vaccine for the deadly acquired immune deficiency syndrome (AIDS). It will come as no surprise then, if medical practitioners fail to consider the possibility of the more common sexually transmitted diseases (STDs) like syphilis as a differential diagnosis in their patients. In November 1989, a six-week-old infant was referred to us with an uncommon haematological complication of early congenital syphilis. This prompted us to look at our figures for congenital syphilis and its mode of presentation. At the same time we had hoped to create an awareness among medical practitioners that this treatable but, more importantly, preventable disease of infancy is still very much with us. Most of us are familiar with the classical skin lesions described in textbooks while forgetting the more uncommon but well known manifestations of this disease, which seems to be on the rise in incidence in many western countries.

PATIENTS AND METHOD

Admissions to the Paediatric Unit of the University Hospital, Kuala Lumpur, over a 10-year period from 1980 to 1989 were reviewed. The records of patients admitted with a diagnosis of congenital syphilis were recalled and studied in detail. The diagnosis of congenital syphilis was based on the following criteria:

1. A positive standard serological test for syphilis (STS) using either the Venereal Diseases Research Laboratory test

(VDRL) or the Rapid Plasma Reagin test (RPR) on the patient's serum and confirmed by a treponemal antibody test using the *Treponema pallidum* haemagglutination test (TPHA).

2. Radiological changes in the long bones, which include periostitis, metaphysitis, osteochondritis; occurring either alone or in combination.

The presence of both of the above together with clinical features compatible with congenital syphilis were considered diagnostic of the disease. The criteria used parallel that recommended by the Centers for Disease Control (CDC), Atlanta⁽¹⁾.

A total of 33 cases were diagnosed as congenital syphilis at the time of admission. Out of this number, only 13 cases fulfilled the above criteria. The other 20 cases were subsequently found to have conditions other than congenital syphilis.

RESULTS

Over a 10 year period between 1980 to the end of 1989, a total of 13 patients with a diagnosis compatible with early congenital syphilis were seen at the University Hospital in Kuala Lumpur. This gives an average of 1 to 2 cases per year. As can be seen, all with the exception of one (Case 1), were aged 3-months and below (Table I). There were four newborns with this condition; two of them were born premature at 30 and 34 weeks gestation respectively. Six of the patients were born with low birthweight (less than 2.5 kg). Although congenital syphilis does not have a predilection for any particular sex, the majority of our patients were males with a male to female ratio of 9:4.

Maternal age ranged from 16 to 32 years (mean 19.9 years) with two primigravidas. There were no unmarried mothers in this study.

There were four mortalities but perhaps only one could be ascribed to be directly related to the underlying syphilis. Patient 4 was referred by a private practitioner with a diagnosis of septicaemia and osteomyelitis of the phalanges of both hands. He developed cardiorespiratory arrest a few hours after admission and succumbed two days later. Patient 8 had birth asphyxia and developed necrotising enterocolitis. He succumbed two months later following a massive gut resection. The other two were premature infants and died from complications related to prematurity.

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Table I
Demographic data on 13 patients with early congenital syphilis

Patient	Age	Sex	Race	Birthweight (kg)	Outcome
1	1 year	male	Chinese	4.30	Survive
2	2 month	male	Chinese	2.00	Survive
3	3 month	male	Chinese	2.20	Survive
4	2 month	male	Indian	NA	Died
5	2 month	female	Indian	NA	Survive
6	6 week	female	Malay	2.40	Survive
7	1 month	male	Indian	2.75	Survive
8	Newborn	male	Indian	1.86	Died
9	Newborn	female	Indian	3.20	Survive
10	Newborn*	male	Malay	1.80	Died
11	Newborn*	female	Malay	2.30	Died
12	3 month	male	Chinese	NA	Survive
13	6 week	male	Indian	2.60	Survive

* = Premature

NA = Not available

The main clinical and laboratory findings in the 13 patients are summarised in Tables II and III.

Table II
Frequency of main clinical manifestations

Presentation	No. of cases	Patient
Pseudoparalysis of a limb	1	1
Maculopapular skin rash	5	2,3,6,7,9
Vesiculobullous eruption	5	5,8,10,11,12
Dactylitis and fever	1	4
Haemolytic anaemia	1	13

Table III
Clinical, laboratory and radiological findings

Findings	No. of cases
Hepatomegaly	11/13
Splenomegaly	13/13
Pallor/documented anaemia (Hb < 12 g/dl)	8/13
Jaundice	1/13
Radiological changes in long bones	9/9
Positive VDRL test on CSF	3/10
Elevated serum immunoglobulin M	8/9

DISCUSSION

Congenital syphilis has been known to man as early as the 15th century. Transplacental transmission from an asymptomatic infected mother to an infant was first described in 1906. In recent years, countries like the United States and the United Kingdom have reported an increase in the number of cases of congenital syphilis^(2,3). In Malaysia, there are no data available on the incidence of congenital syphilis. There is, however, an increase in the incidence of infectious syphilis in the adult population in Malaysia over the years from 1983 to 1987 (Table IV). It can be seen that the incidence of notified syphilis has increased from 3.89 per 100,000 population in 1983 to 11.41 per 100,000 in 1987. Bearing in mind the notoriously low rate

Table IV
Incidence of infectious syphilis in Malaysia

Year	Cases reported	Incidence/100,000 population
1983	556	3.89
1984	849	5.56
1985	928	5.91
1986	1440	8.93
1987	1887	11.41

Source : Annual Report 1986 and 1987
Ministry of Health, Malaysia

of notification for infectious diseases, the actual figures quoted are certainly only the tip of the iceberg.

Congenital syphilis is a totally preventable and treatable disease. It will continue to exist as long as infectious syphilis is not eradicated in the young adults.

Early congenital syphilis is that in which clinical manifestations appear within the first two years of life while those which appear after this time are designated late congenital syphilis. The disease often involves multiple body organ systems and thus can be easily confused with many other diseases of the newborn. Clinical signs are usually absent at birth and may be non-specific (eg. pallor, skin rash and hepatosplenomegaly) while late symptoms and signs are often diverse. Three of the newborns had the typical vesiculobullous eruption on the palms and soles while the fourth developed a maculopapular skin rash over the palms and soles a few days after birth.

Severe haematological disturbances in congenital syphilis have been well established^(4,5). Patient 13 presented with acute haemolysis which was initially thought to be due to erythrocytic G6PD deficiency, a condition which is common in this part of the world. The diagnosis of early congenital syphilis was made only when the typical maculopapular rash appeared on the palms and soles on the third hospital day.

The diagnosis of early congenital syphilis thus requires a high degree of awareness and vigilance. Stevens (1987) remarked that "the chief pitfall in the diagnosis of early infection is the paediatricians' failure to include it among the differential diagnoses of a large number of illnesses in infancy"⁽⁶⁾.

Splenomegaly appears to be the only consistent physical sign, with or without an associated hepatomegaly.

It is rare for congenital syphilis to present with musculo-skeletal complaints alone⁽⁶⁾. We found one patient who presented with pseudoparalysis of the lower limbs while another had severe dactylitis of the fingers. On the other hand, radiological changes in the long bones are common and are present in all those cases where an X-ray of the long bones was carried out.

Although the non-treponemal VDRL test is positive in all our cases, diagnosis of congenital syphilis is difficult to make on the basis of serological tests alone as its presence may be due to passive antibody acquired from the mother which again may be present non-specifically during pregnancy. It is useful as a screening test for suspected congenital and acquired syphilis and as quantitative serological test to assess disease activity and efficacy of therapy⁽⁷⁾. However, coupled with the elevated serum immunoglobulin M level (which is the case in all our patients where it was carried out) plus the clinical signs and radiological changes, a presumptive diagnosis of early congenital syphilis may be made. This could then be confirmed with the more specific TPHA test.

The significance of a positive VDRL test on the CSF is

debatable⁽⁸⁾. The CDC in its diagnostic classification of congenital syphilis regards a positive VDRL in the CSF indicative of a compatible case of congenital syphilis neurological affliction and recommends treatment with at least a 10-day course of crystalline penicillin.

Persistent intrauterine infection may result in death in about 25% of cases; another 25-30% may die shortly after birth⁽⁹⁾. Steps must be taken to identify and treat infected mothers so as to prevent this disease. Routine antenatal serological tests must be carried out early in pregnancy and repeated, if necessary, in the third trimester. An early negative test may mean either that the mother is incubating syphilis or she may have acquired it later in pregnancy. If a mother is found to have syphilis, congenital syphilis will invariably be prevented if treatment is completed at least 2 weeks before delivery⁽¹⁰⁾.

Preventable factors would include providing adequate and appropriate prenatal care to expectant mothers and continued community surveillance and contact tracing.

SUMMARY

A retrospective study on the clinical, laboratory and radiological presentation of 13 consecutive cases of congenital syphilis was carried out over a 10 year period at the University Hospital, Kuala Lumpur. All the cases were less than two years old at presentation. Skin lesions either in the form of a non-specific erythematous maculopapular rash or the typical vesiculobullous eruption over the palms and soles are the most common presentation.

Splenomegaly is the most consistent clinical sign followed by hepatomegaly and pallor. A presumptive diagnosis of early congenital syphilis is made if the patient's VDRL test is positive in the presence of the above clinical features. The serum immunoglobulin M level is a useful screening test for the presence of active infection and radiological changes in the long bones are universally present in those cases where an X-

ray of the long bones was carried out.

Early congenital syphilis could and should be prevented. The result of the non-treponemal VDRL test carried out during the antenatal visits should be made available early so that treatment could be completed at least two weeks before delivery. Cases of infectious syphilis in the young adults must be notified promptly and contact tracing carried out so that they can be treated as well in an effort to break the chain of transmission of the disease. Finally, we should also be aware of the other less common modes of presentation of early congenital syphilis so that early and appropriate treatment could be given.

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