

Cleft lip as a presentation of congenital syphilis

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Abstract

Congenital syphilis may present with unusual symptoms in early stages which needs to be identified for prompt treatment. Here, we present a case of 13-day-old female child with congenital syphilis presenting with cleft lip.

Key words: Cleft lip, congenital syphilis, venereal disease research laboratory

INTRODUCTION

Syphilis, a debilitating illness, has been a global problem since ages. In the third-world countries, 3–15% of the women are infected with syphilis in their reproductive age.^[1,2] As per the World Health Organization (WHO), about 2.1 million pregnant women are diagnosed to have active syphilis each year. Most of them are young females without adequate antenatal care, having 95% chance of transferring the infection to their fetus transplacentally. The WHO estimates that every year, maternal syphilis is responsible for 460,000 abortions or still birth, 2,70,000 congenital syphilis cases, and the birth of 2,70,000 premature or low birth babies.^[3]

Besides abortions, still births, and perinatal deaths, live born babies may have early (before 2 years of age) or late (after 2 years) manifestations of congenital syphilis. Two third of these cases may be asymptomatic at birth and develop malformations of teeth and skeleton later in life.

Here, we report a case of a 13-day-old female child

with a cleft lip as a presenting feature of congenital syphilis.

CASE REPORT

A 13-day-old female child with a cleft lip was referred from Department of Plastic Surgery to Skin and Venereal Disease OPD, Medical College, Vadodara. On history, it was found that the baby was delivered at home and the mother had not taken any antenatal visits. Further as told by mother herself, she did not have any illness during her pregnancy nor did she consume any drugs. Father of the baby did not have any history of any sexually transmitted diseases (STDs) but had high risk behavior. The baby had two siblings; all of them were healthy without any congenital anomalies.

On examination, it was found that the baby had upper cleft lip with no palatal involvement (Figure 1). There were no apparent bony abnormalities. No abnormality was detected during general and systemic examinations. There was no clinical evidence of STD in parents. Based on the high risk behavior of father, an attempt was made to rule out syphilis. The parents were subjected to Serum Venereal Disease Research Laboratory (VDRL) testing and were found to be positive with reactivity of mother and father at 1:32 and 1:16 dilutions, respectively. The baby was subjected to a series of investigations. The baby was found positive for VDRL (reactive at 1:4 dilutions). On CSF examination of the baby, the VDRL was nonreactive

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Figure 1: A 13-day-old female child with cleft lip

with CSF proteins 60 mg/dl, fitting into presumptive diagnosis of syphilis. HIV test of the parents was nonreactive. Other laboratory investigations (Complete blood count, platelet count, etc.) were within normal limits. X-ray of the limbs was normal.

The infant received injection benzathine penicillin G 50,000 units/kg/dose intramuscularly (IM) in a single dose. Parents were also treated simultaneously with injection benzathine penicillin G 2.4 million units in single dose.

The infant's VDRL titres became 1:2 by 3 months and undetectable by 6 months of age.

DISCUSSION

The case definition of congenital syphilis as per Centers for Disease Control (CDC), USA, includes newborns with clinical evidence of active syphilis as well as those who do not present any signs and are born to the mothers with untreated or inadequately treated syphilis.^[4]

About 60% of infants born with congenital syphilis are asymptomatic at birth. Symptoms develop within the first 2 months of life.^[5] Early congenital syphilis usually manifest as rhinitis, vesicular and other skin eruptions, bone abnormalities, chorioretinitis, hepatosplenomegaly, and nephrotic syndrome. Late congenital syphilis, which presents after 2 years of age, may have a variety of skeletal and dental defects, interstitial keratitis, and eighth nerve deafness (Hutchinson's Triad).^[6] Early identification and treatment of syphilis will help to prevent these outcomes.

The causes of orofacial clefts among most infants are unknown. Cleft lip and cleft palate are thought to be caused by a combination of genes and other factors such as exposure to infections like rubella, syphilis, and chickenpox; drugs like antiepileptics, oral contraceptive pills, isotretinoin; maternal diet

(folic acid deficiency); or ionizing radiations (X-rays). In a study, in Latin America it was found that acute (influenza) and chronic (epilepsy and syphilis) maternal illness during first trimester of pregnancy and gravidity higher than four were risk factors for cleft lip.^[7]

Due to limited resources a presumptive diagnosis of congenital syphilis is frequently the basis for treatment of newborn. And the presumption of transmission is raised by the positive serology of the mother with inadequate antenatal care. Treatment is based upon a maternal positive nonspecific treponemal test at pregnancy or delivery in combination or not with clinical symptoms of newborn. However, confirmation of syphilis requires demonstration of syphilis spirochete in the tissues or body fluids by detection of anti-treponemal IgM antibodies or by DNA or RNA amplification, which could not be done in this case. Hence, a presumptive diagnosis of syphilis was made as per CDC criteria.

CONCLUSION

Despite comprehensive antenatal screening recommendations and inexpensive treatment, congenital syphilis has long been and continues to be a public health concern, causing substantial morbidity and adverse outcomes. Screening of pregnant women for syphilis and treatment with penicillin is an effective measure to prevent congenital syphilis. Rare associations and manifestations of congenital syphilis need to be identified to prevent adverse outcome.

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