Late Congenital Syphilis in a Pediatric Patient

ABSTRACT

Syphilis is an infectious disease which is on rise these days due to various etiological factors, like HIV, intravenous drug usages, promiscuous sex. It is broadly of three types—primary, secondary and tertiary. Congenital syphilis comes under tertiary type. This case report describes about female child having late congenital syphilis infection. Congenital syphilis occurs due to the transmission of the disease from an infected mother to the unborn infant through the placenta. The child had symptomatic congenital syphilis with dental manifestations.

Keywords: Child, Dental teeth, Syphilis.

INTRODUCTION

As we have ventured in 21st century, there is an upsurge of so many new types of infectious disease, affecting human race. Among these, bacterial infection syphilis which was long forgotten disease affects the pregnant women even today resulting in prenatal morbidity and mortality.1

Although worldwide incidence of syphilis decreased over the last 25 years and the disease was on the verge of being eliminated in many countries in 1998, infection rates have recently increased.2 According to the centers for disease control and prevention (CDC), a total of 49,903 cases of syphilis were reported in the United States in 2012.2

Syphilis is caused by Treponema pallidum, a spirochete that cannot survive for long outside the human body. Treponema pallidum enters through the mucous membranes or skin, reaches the regional lymph nodes within hours, and rapidly spreads throughout the body.5 Infection is usually transmitted by sexual contact (including genital, orogenital and anogenital) but may be transmitted nonsexually by skin contact or transplacentally.3 Risk of transmission is about 30% from a single sexual encounter with a person who has primary syphilis and 60 to 80% from an infected mother to a fetus. Infection does not lead to immunity against reinfection.5

Congenital syphilis is a severe, disabling infection often with grave consequences. Approximately, 66% of infected infants are asymptomatic at the time of birth and are identified in routine prenatal screening.6-8 World Health Organization (WHO) estimates that 2 million get infected with syphilis every year. Intrauterine infection with spirochetes T. pallidum can result in still birth, hydrops fetalis, or preterm birth or be asymptomatic at birth.9,10

Questions regarding prevention and management of congenital syphilis persists because the diagnosis of the suspected cases and management may be confusing, and the potential for severe disability is high when cases are missed. The venereal disease research laboratory (VDRL) test and rapid plasma reagin (RPR) are used to detect the antibody called reagin, which is produced by the immune system’s response to T. pallidum infection. ‘False negative’ results may occur when these tests are performed during the first 3 to 6 weeks following infection (primary syphilis); negative results do not rule out syphilis during this time. The continuing prevalence of the disease reveals the failure of control measures established for its prevention.

We put forth a case of symptomatic congenital syphilis with dental manifestations. The report stresses upon the importance of implementing WHO recommendation that all pregnant women should be screened for syphilis in the first antenatal visit.

CASE REPORT

An 11-year-old female patient (Figs 1 to 3) reported to the department of pedodontics and preventive dentistry with a chief complaint of discolored upper front and lower back teeth. As told by the grandmother, the child was adopted by her as she had found the child on road side.
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As told by patient’s grandmother, child has habit of scratching her gingival sulcus, both in upper and lower arches during daytime. The child’s grandmother gave the history of rash and mucocutaneous lesions. The child did not have any snuffles or feeding difficulty. The child’s grandmother complained that child has continuous pain in her joints and self-limiting swellings in all joints.

Extraoral examination showed saddle shape nose. Anterior fontanel was flat, and there was no asymmetry of movements or cranial nerve palsy. On intraoral examination, screw driver shaped incisors were present wrt 11 and 21 (Figs 4 and 5). Mulberry molars were present wrt 36 and 46 (Figs 6 and 7). Incisors were screw driver’s shape discolored with notching (Hutchinson’s incisor). Occlusal caries were present wrt 36, 46 and 55.

Gingiva was inflamed. Generalized stains were present.

After intraoral examination, the provisional diagnosis of congenital syphilis was made, taking into consideration of amelogenesis imperfecta, septic arthritis and traumatic arthritis as differential diagnosis.

Patient underwent routine blood investigation, VDRL test, eye and ear examination, neurological examination and skeletal examination. Ultrasound examination of abdomen and pelvis showed no abnormality. Leg anteroposterior X-ray was normal. There was no lymphadenopathy and no apparent bony abnormality. There was no evidence of neurological or cardiovascular involvement.

Investigations revealed normal blood picture and C-reactive protein. Neurological, cardiovascular ENT and ophthalmological examinations were normal. Venereal disease research laboratory result showed 1:4 dilution and was reactive (Fig. 8).

Based on reports of above-mentioned investigations, confirmatory diagnosis of congenital syphilis was made.

DISCUSSION

In 21st century when congenital syphilis is thought to be eradicated, it is re-emerging due to pitfalls in ANC or screening. One of the most striking features of congenital syphilis is that it may be transmitted from infected
parents to their offspring, the children exhibiting the manifestations that characterize the acquired form of the disease.

The more recent the syphilis in the parent, the greater is the risk of the disease being communicated to the offspring; so that if either parent suffers from secondary syphilis the infection is almost inevitably transmitted.

In contrast to the acquired form, inherited syphilis is remarkable for the absence of any primary stage, the infection being a general one from the outset. The spirochete is demonstrated in incredible numbers in the liver, spleen, lung and other organs, and in the nasal secretion, and, from any of these, successful inoculations in monkeys can readily be made. The manifestations differ in degree rather than in kind from those of the acquired disease; the difference is partly due to the fact that the virus is attacking developing instead of fully formed tissues.

Primary syphilis, the earliest stage, is characterized by the presence of lesions at the site of *T. pallidum* entry and subsequent development of regional lymphadenopathy. In secondary syphilis, which occurs about 2 to 10 weeks after infection hematogenous dissemination of *T. pallidum* causes several systemic findings, including fever, malaise, generalized lymphadenopathy and mucocutaneous lesions (papular, macular, annular or follicular lesions). The signs and symptoms of primary and secondary syphilis resolve spontaneously and patients then enter the latent stage of infection (Anderson et al, 1989; Hutchinson and Hook, 1990). After a variable period of latency, tertiary or late stage disease develops in about one third of untreated patients. Manifestations may take up to 10 years to appear and then present themselves as benign tertiary (gummatous lesions), cardiovascular syphilis, or neurosyphilis (Hook and Marra 1992). Conclusive diagnosis of syphilis infection is based on confirmation of the clinical signs and symptoms with laboratory tests (Hook et al).

Syphilis has wide systemic and oral manifestations. But in our patient we saw it has affected the bone and oral cavity. Literature review shows its effect on bone and oral cavity as:

**Affections of the Bones**

Swellings at the ends of the long bones, due to inflammation at the epiphyseal junctions, are most often observed at the upper end of the humerus and in the bones in the region of the elbow.

**Changes in the Permanent Teeth**

These affect specially the upper central incisors, which were dwarfed and stand somewhat apart in the gum, with

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**Fig. 6:** Occlusal view (max)

**Fig. 7:** Occlusal view (mand)

**Fig. 8:** Report of the patient
their free edges converging toward one another. They were tapering or peg-shaped, and present at their cutting margin a deep semilunar notch. These appearances are commonly associated with the name of Hutchinson, who first described them. Affecting as they do the permanent teeth, they are not available for diagnosis until the child is over 8 years of age. Henry Moon drew attention to a change in the first molars; these are reduced in size and dome-shaped through dwarfing of the central tubercle of each cusp. In our case also we made a provisional diagnosis based on intraoral examination which revealed characteristic Hutchinson’s incisors and Mulberry molars.

While a considerable number of syphilitic children grow-up without showing any trace of their syphilitic inheritance, the majority retain throughout life one or more of the following characteristics, which may therefore be described as permanent signs of the inherited disease: Dwarfing of stature from interference with growth at the epiphyseal junctions; the forehead low and vertical, and the parietal and frontal eminences unduly prominent; the bridge of the nose sunken and rounded; radiating scars at the angles of the mouth; perforation or destruction of the hard palate; Hutchinson’s teeth; opacities of the cornea from antecedent keratitis; alterations in the fundus oculi from choroiditis; deafness, depressed scars or nodes on the bones from previous gummatas; ‘sabre-blade’ or other deformity of the tibia.

This article highlights the awareness of the importance of dental findings in systemic disease of late congenital syphilis, which is a very rare clinical entity and its importance to make early diagnosis, and its proper management can prevent further complications. Although an effective treatment is available since the introduction of penicillin in the mid-20th century, it still remains as an important public health problem.

CONCLUSION
Prevention of Congenital Syphilis

- Ensure that official public health statutes and/or regulations mandate STS on all pregnant women at the time of the initial prenatal visit and early in the third trimester.
- Encourage prenatal screening for syphilis wherever pregnant women are seen for healthcare, including women, infants and children (WIC) programs, methadone maintenance clinics, detention facilities, and prenatal care facilities; whenever possible, review existing clinic protocols and suggest specific amendments to the clinic medical director.
- Conduct selective serologic screening of women of childbearing age in groups with an increased risk of infection, e.g. women residing in neighborhoods that have a particularly high incidence of syphilis.
- Deliver educational messages to the medical community about laboratory tests, diagnostic criteria, treatment, and follow-up of patients who are at risk of infection and who may be pregnant.
- Develop and disseminate public service educational messages to women who share demographic characteristics with the women most often diagnosed with early syphilis.

Regardless of the stage of pregnancy, patients who are not allergic to penicillin should be treated with penicillin according to the dosage schedules appropriate for the stage of syphilis as recommended for nonpregnant patients. Recommended regimens for symptomatic or asymptomatic infant: aqueous crystalline penicillin G 50,000 units/kg IM or IV daily in two divided doses for a minimum of 10 days or Aqueous procaine penicillin G 50,000 units/kg IM daily for a minimum of 10 days.

For asymptomatic infants whose mothers were treated adequately with a penicillin regimen during pregnancy but whose follow-up cannot be ensured, many consultants recommend treatment with benzathine penicillin 50,000 units/kg IM in a single dose. Data on the efficacy of this regimen in congenital neurosyphilis are lacking; therefore, if neurosyphilis cannot be excluded, the 10-day regimens of aqueous crystalline penicillin or procaine penicillin are recommended. Only penicillin regimens are recommended for neonatal congenital syphilis.

All patients with early syphilis or congenital syphilis should be encouraged to return for repeat quantitative nontreponemal tests at least 3, 6 and 12 months after treatment.

Syphilis is in rise among adult and pediatric population due to varied etiological factors. In this case, in spite of repeated medical visit child went undiagnosed for late congenital syphilis which may be due to improper documentations and missing out on small findings. Understanding the pathophysiology and progression of this disease still remains challenge even in the modern era.

REFERENCES
1. South East Asia and world health organisation, regional strategy for the elimination of congenital syphilis, WHO, New Delhi, India, 2009.


