An inscrutable entity: A case report of late congenital syphilis

Pratiksha Kanubhai Rathwa, Jatin Chauhan, Nidhi Pravinkumar Patel, Hiral Shah, Nipul V Vara
Department of Dermatology, Venereology and Leprosy, Government Medical College and S.S.G. Hospital, Vadodara, Gujarat, India

Address for correspondence:
Dr. Pratiksha Kanubhai Rathwa, Department of Dermatology, Venereology and Leprosy, Government Medical College and S.S.G. Hospital, Vadodara, Gujarat, India.
E-mail: rathwa.pratiksha.rpk@gmail.com

Abstract

Congenital syphilis is an outcome of maternal syphilis that occurs due to the transmission of *Treponema pallidum* via the placenta of untreated or inadequately treated pregnant women to their newborns. It is now a very rare cause of neurological, developmental, and musculoskeletal disability and death in infants after the advent of penicillin. Here, we report a case of late congenital syphilis presented with classic stigmata of syphilis at the age of 10 years. Reactive serological titer of rapid plasma reagin test and venereal disease research laboratory test confirmed the diagnosis.

Key words: Congenital syphilis, crystalline penicillin G, rapid plasma reagin, *Treponema pallidum*

Introduction

Congenital syphilis ravaged the humankind for almost 350 years. There was dramatic decline in the incidence following the introduction of penicillin in 1943. However, the upsurge in the cases of congenital syphilis began in developing countries despite advent of penicillin which is useful to treat syphilis since the early 1950s.[1] The incidence of congenital syphilis in childhood sexually transmitted diseases is < 1/1000 in Indian patients.[2] This highlights the inadequate prenatal care as the leading predisposing factor. This underscores the importance of timely antenatal and postnatal screening and treatment of seropositive mothers and their babies with penicillin.

Case Report

A 10-year-old female child was referred to us from the pediatric department for an asymptomatic erythematous ruptured nodular lesion below the chin and depressed scar over the forehead and forearm for the past 2–3 months. Along with these cutaneous features, she also had complaints of headache, vomiting, and one episode of seizure on arrival. On clinical examination of the child, frontal bossing [Figure 1a] and depressed scar over the left side of the forehead and left forearm suggestive of "healed gummata" [Figure 2a] were noted. A single ruptured subcutaneous nodule with pus discharge was noted below chin suggesting active gumma [Figure 1b]. Examination of the oral cavity showed peg-shaped upper central incisor teeth (Hutchinson teeth) and high arched palate [Figure 1c]. Oral mucosa was normal. Clinical examination of the extremities revealed limb deformity of the left forearm and "Sabre shin deformity" in the right leg [Figure 2c]. With these clinical features and history of the patient, congenital syphilis and tuberculous meningoencephalitis were kept as differential diagnoses. Abnormal anterior bowing with cortical thickening and mild periosteal reaction were noted along the right tibial shaft [Figure 2d] on radiological examination of the tibia, and X-ray of the left wrist showed mixed osteolytic sclerotic lesion at the left distal metaphyseal radius, suggesting chronic osteoperiostitis in affected bones [Figure 2b]. There were absence of any ocular findings and no neural deafness detected on audiometry. Magnetic resonance imaging of the brain was done which showed evidence of granulomatous lesion or meningioma. Cerebrospinal fluid (CSF) examination revealed raised protein, and venereal disease research laboratory test for CSF came out to be reactive. Mantoux test and HIV were negative. Treponema Pallidum Haemagglutination Assay test was positive, and rapid plasma reagin (RPR) titer of the patient was >1:256, thus confirming late congenital syphilis. The patient was living with her grandparents, and thus, serological tests for syphilis could not be performed in her parents and her siblings as they are no longer living together with her. The gestational history of the patient suggested her birth as a result of nonconsanguineous marriage, and she was born by normal vaginal home delivery with no history of antenatal

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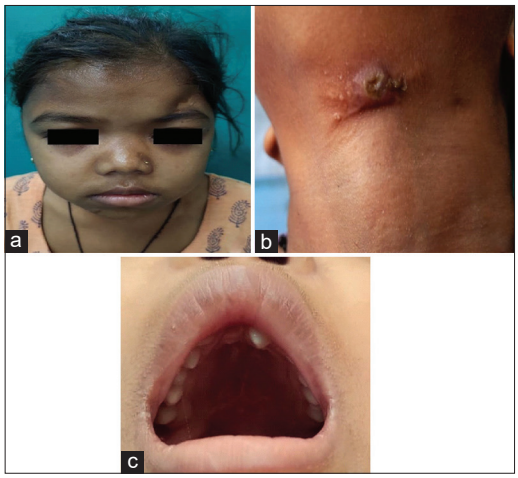

Figure 1: (a) Frontal bossing, saddle nose deformity and healed gummata over the forehead, (b) active gumma below chin, (c) Hutchinson teeth with high arched palate

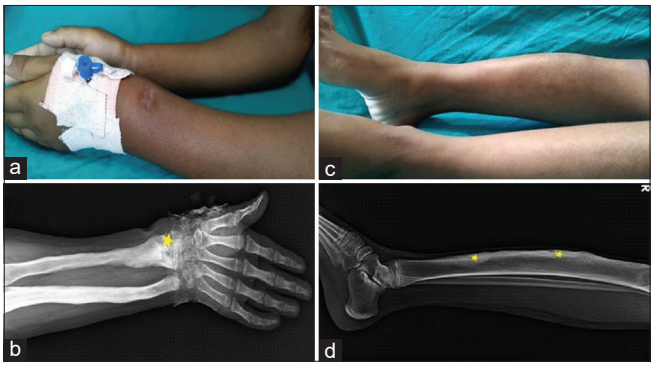

Figure 2: (a) Healed gummata and deformity of left forearm, (b) osteolytic sclerotic lesion in X-ray of the left wrist, (c) Sabre shin deformity, (d) anterior bowing with cortical thickening, mild periosteal reaction in X-ray of the right lower limb

care taken in mother or any history of postnatal screening of patient at the time of delivery. There was no history of pregnancy loss in mother during previous pregnancy. Thus, reactive serological tests with significant raised titer of RPR in patient with classic clinical examination findings were pointing toward the diagnosis of late congenital syphilis. The patient was started injection aqueous crystalline penicillin G 900,000 IU IV every 12 hourly for 7 days and every 8 hourly thereafter for a total of 10 days.

Discussion

Congenital syphilis is an outcome of maternal syphilis which occurs due to the transmission of *Treponema pallidum* via the placenta of untreated or inadequately treated pregnant women to their newborns. In the postpenicillin era, it has become a rare cause of neurological, developmental, and musculoskeletal disability and death in infants. The clinical course of congenital syphilis can arbitrarily divided into: (1) early congenital syphilis: features typically appear within the first 2 years of life and (2) late congenital syphilis: features that appear after 2 years of life. The early stage goes unnoticed in approximately 80% of children which are ultimately diagnosed as having late congenital syphilis. Late congenital syphilis in the child corresponds to tertiary syphilis in the adults. Inadvertent use of antibiotics for other intercurrent infections has modified the expression of the disease to such an extent that classic syndromes of late congenital syphilis are now rare.[3] The clinical manifestations are either stigmata or hypersensitivity or inflammatory reactions and involve multiple systems. The stigmata of late congenital syphilis represent the delayed consequences of the localized inflammatory processes established at the sites of treponemal infection during the early stage of disease, which makes the early identification and prompt treatment of infants born to seropositive mothers crucial. The main characteristics of late congenital syphilis include stigmata presenting as frontal and parietal bossing, saddle nose deformity, Hutchinson teeth, high arched palate, Sabre shin deformity, interstitial keratitis, gummata, neural deafness, and clutton's joint. Also noteworthy is the sequelae of osteoperiostitis, which is a characteristic radiological finding (as present in our patient). The cornerstone of congenital syphilis control in the present times is antenatal screening and timely appropriate treatment of seropositive mothers with penicillin. With vigilant focus on maternal and child health programs, congenital syphilis is preventable, but it continues as a scourge in the face of declining seropositivity. This case report of a late congenital syphilis is a tragic reflection of pitfalls in the antenatal screening of pregnant mothers and postnatal screening of babies soon after birth which cannot be achieved with perfection, due to patient's ignorance and illiteracy as in our case. Thus, emphasis needs to be put on implementation of prevention strategies such as raising awareness among people, and early identification and prompt treatment modalities for maternal and neonatal syphilis is the key to avert significant future morbidity.

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Conflicts of interest

There are no conflicts of interest.

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