An infant with acral peeling of skin: A curious case of congenital syphilis

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Abstract

Congenital syphilis (CS) is a vertically transmitted infection caused by the spirochete *Treponema pallidum*. It is seen rarely due to proper antenatal screening. Signs and symptoms appear within the first 2 years of life in early CS and after 2 years in late CS. Failure to diagnose and treat CS in its early stages can result in higher morbidity and mortality. Skin manifestations can guide toward the diagnosis of CS at an early stage. Here, we report a 2-day-old neonate who presented with acral peeling of skin along with respiratory distress and hepatosplenomegaly. Clinical suspicion of CS was made and subsequently confirmed by a positive venereal disease research laboratory test in both mother and child. The child was treated with aqueous crystalline penicillin G as per the CDC guidelines.

Key words: Acral peeling of skin, aqueous penicillin, congenital syphilis, hepatosplenomegaly, respiratory distress

Introduction

Congenital syphilis (CS) is a rare but potentially fatal condition that occurs because of vertical transmission of *Treponema pallidum* from the mother to the fetus. It is completely preventable through proper antenatal screening and treating infected mothers with penicillin. Delay in diagnosis and treatment can result in infant mortality and various sequels in survivors, resulting in lifelong morbidity. Here, we report a 2-day-old neonate who presented with peeling of acral skin with respiratory distress and was later diagnosed as CS.

Case Report

A 2-day-old newborn baby born out of a nonconsanguineous parentage was admitted to the neonatal intensive care unit with respiratory distress. The birth weight was 2.78 kg. There was a history of thick meconium aspiration with a delayed cry. The baby cried after 2 min following the bag and mask ventilation. There was hepatosplenomegaly, with the liver being 7 cm and the spleen 4 cm, respectively. Investigations revealed total bilirubin 6.7 mg/dL (direct bilirubin 3.9 and indirect bilirubin 2.9), serum glutamic-oxaloacetic transaminase - 707 U/L, serum glutamic-pyruvic transaminase - 1574 U/L, alkaline phosphatase - 440 U/L, platelet - 80,000/cmm, total leukocyte count - 21,350/cmm, and C-reactive protein - 111.7. Urine and blood cultures were sterile. Chest X-ray was normal. Because of the presence of hepatosplenomegaly, the baby was thoroughly investigated for toxoplasmosis, hepatitis B, rubella, cytomegalovirus, herpes simplex infection and tuberculosis, which came out to be negative. On the 6th day, the baby developed peeling of the skin from the acral parts and trunk, for which dermatology consultation was sought. The mother gave a history of erythematous rash over the trunk and extremities on the 2nd day of birth. On dermatological examination, there was peeling of the skin from the palms and soles [Figure 1a and b]. Based on skin peeling from the acral areas, physiological desquamation of neonatal skin, continual peeling skin syndrome, and CS were kept as differentials. The patient was subsequently investigated. Venereal disease research laboratory (VDRL) titer of the mother and the baby came out as 1:4 and child 1:16,

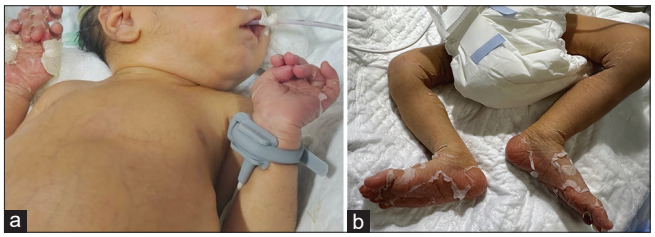

Figure 1: (a) Peeling of the skin from the palms with abdominal distension. (b) Peeling of skin from lower extremities and soles

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respectively. Cerebrospinal fluid (CSF) analysis showed glucose 37 mmol/L, protein 191.92 mg/dL, cell count 15 cells/mm^3, and lymphocytes 100% and CSF VDRL came out to be negative. The patient was finally diagnosed with CS based on clinical findings and laboratory reports and was started with aqueous crystalline penicillin G 50,000 IU/kg/day intravenously (IV) 12 hourly in the first 7 days of life, followed by 50,000 IU/kg per dose IV every 8 hourly for the next 3 days to complete a total 10 days of treatment.

Discussion

CS is caused by *T. pallidum*, which is transmitted vertically *in utero* or during intrapartum from the mother. The likelihood of transmission from the mother to the child is directly related to the stage of pregnancy at which the infection is acquired and the stage of syphilis. The probability of transmission is 70% to 100% during primary syphilis, 40% during early latent syphilis, and 10% in late latent syphilis.[1] Pregnant women should be screened for syphilis as per the NACO guidelines at their first antenatal checkup visit (preferably in the first trimester) for early detection. CS is a potentially fatal but preventable disease. Clinical symptoms appear within 2 years in case of early CS and after 2 years in case of late CS.[2]

Table 1: Clinical features of early congenital syphilis[2,3]

Early CS - infectious (first 2 years)
Mucocutaneous
Rhinitis
Maculopapular rash resolving with desquamation and crusting
Pemphigus syphilitics (vesiculobullous lesion)
Condylomata lata (perioral and perianal)
Hair and nails
Patchy alopecia with brittle and sparse hair
Syphilitic paronychia
Hematology
Anemia
Thrombocytopenia and
Petechia
Reticuloendothelial
Hepatosplenomegaly
Palpable epitrochlear lymphadenopathy
Radiology
Pseudoparalysis of parrot[3]
Wimberger's sign (demineralization and destruction of the proximal tibial metaphysis)
Eyes
Chorioretinitis
Cataract
Glaucoma
Neurological
Aseptic meningitis
Cranial nerve palsies
Seizure
GIT
Rectal bleeding
Malabsorption
Necrotizing enterocolitis
Other
Myocarditis, pneumonia
Pancreatitis, nephrotic syndrome

CS=Congenital syphilis; GIT=Gastrointestinal tract

Diagnosis of CS can be difficult because more than two-thirds of affected infants are asymptomatic at birth, and signs of symptomatic infants may be nonspecific or subtle. On top of this, some affected infants may have atypical presentations. Familiarity with the diverse presentations is essential to diagnosis. Delayed diagnosis and treatment can lead to late, persistent clinical features of intellectual disability, skin gummas, scarring, hearing deficits, and skeletal abnormalities. The detailed cutaneous and systemic manifestations of early and late CS are illustrated in Tables 1 and 2. Our case had respiratory distress because of hepatosplenomegaly and the only skin manifestation was peeling of skin from extremities.

Desquamation of the skin found in early CS can be confused with other conditions like physiological desquamation of neonatal skin and continual peeling skin syndrome. Desquamation of neonatal skin has been reported in 65%–75% of normal newborns. Scaling is confined to ankles, hands, and feet in term infants, whereas postterm infants have generalized scaling along with other cutaneous signs of postterm like the absence of vernix, long nails, and hair, and decreased subcutaneous fat.[5] Peeling skin syndrome is characterized by asymptomatic shedding of the skin in large sheets since birth. In these patients, peeling of the skin can be produced by rubbing the skin. Histopathology can help confirm the diagnosis, in which the separation of the stratum of the corneum from the underlying granular layer is seen.[6] Diagnosis of CS is usually confirmed by non-treponemal tests: VDRL slide test, rapid plasma reagin (RPR) card test, automated reagin test (ART), reagin screen test (RST), and treponemal tests: T. pallidum immobilization test (TPI), Fluorescent treponemal antibody absorption test (FTA-Abs), and FTA-Abs double staining test (FTA-Abs DS), T. pallidum hemagglutination assay (TPHA), microhemagglutination for

Table 2: Clinical features of late congenital syphilis[4]

Late CS - noninfectious (after 2 years)
Teeth
Hutchinson's teeth
Mulberry molars
Eyes
Interstitial keratitis
Ears
Sensory neural hearing loss (8th cranial nerve deafness)
Cutaneous
Rhagades (radiating fissuring at the angle of the mouth)
Gummas (granulomatous inflammatory response to spirochetes)
Central nervous system
Mental retardation, seizures, cranial nerve palsies, hydrocephalus
Skeletal
Saber shins (bowing of the tibia due to the thickening of the middle third)
Higouménakis sign (thickening of the sternal end of the clavicle)
Frontal fibrosing
Saddle nose
Impaired maxillary growth
Clutton joints (painless arthritis of the knees)
Hematology
Paroxysmal cold hemoglobinuria
Hutchinson's triad[4]
Interstitial keratitis
Sensorineural hearing loss, and Hutchinson's teeth

CS=Congenital syphilis

T. pallidum (MHA-TP), and hemagglutination treponemal test for syphilis (HATTS) Enzyme-linked immunosorbent assay (ELISA). Other supportive investigations like dark field microscopy of nasal discharge if present, CSF analysis and CSF polymerase chain reaction to detect Treponemal DNA and imaging (X-ray of chest and long bones) can help in confirming the diagnosis. The neonatal nontreponemal serologic titer is fourfold higher than the mother's titer at the delivery.

The prognosis is excellent if diagnosed early and treated with penicillin.

The curative treatment of a confirmed case of CS includes aqueous crystalline penicillin G 100,000–150,000 IU/kg body weight/day, administered as 50,000 units/kg body weight/dose IV every 12 h during the first 7 days of life and thereafter every 8 h for 3 days to complete a total of 10 days treatment. CDC recommends CSF analysis for VDRL, cell count, and protein, complete blood count, differential and platelet count, and other tests as clinically indicated.[7]

Our case had hepatosplenomegaly and respiratory distress as the main problem. The only positive skin finding on examination was peeling of skin from extremities, which makes the case interesting to report. Hence, the clinicians have to be more suspicious and should have a sound knowledge of varied cutaneous presentations of CS which can guide toward early diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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