ABSTRACT

**Background:** Congenital syphilis is an infectious disease of newborns caused by *Treponema pallidum*, a spirochete bacterium, from vertical transmission from infected mother to fetus. Diagnosis of congenital syphilis can be difficult because two-thirds of affected infants are asymptomatic at birth, and the signs of symptomatic infants are diverse. Familiarity with the various presentations is essential to early diagnosis and treatment.

**Case Description:** A male preterm, 35 weeks gestation baby, born by vaginal delivery from G1P0 mother with syphilis latent phase, with *Treponema pallidum* hemagglutination assay (TPHA) titer reactive (1:2560) with no treatment during the antenatal period. The baby was born with low birth weight, respiratory distress syndrome, hepatosplenomegaly, and skin rash. Diffuse desquamation and fissures, and multiple eroded pale blisters are known as pemphigus syphilitics. TPHA was also reactive at the same level as the mother. Complete blood count showed severe respiratory acidosis (pH: 6.8), leucocytosis (46,420/uL), thrombocytosis (414,000/uL), lymphocytosis (40,3%), and monocytosis (16%). Radiology showed severe atelectatic left lung and course nodular congenital pneumonia of the right lung. The baby was intubated and injected with penicillin procaine 50,000 IU/kg/day intramuscularly in the neonatal intensive care unit (NICU). However, due to severe atelectatic lung and acidosis, this case resulted in a fatal outcome.

**Conclusion:** Broad desquamation, multiple fissures, and eroded pale blisters (vesiculobullous lesions) known as pemphigus syphilitics should alert clinicians as an early manifestation of congenital syphilis. Other manifestations include congenital pneumonia, typically atelectatic or nodular type, and hepatosplenomegaly. Treatment should be started promptly to decrease its morbidity and mortality.

**Keywords:** congenital syphilis, pemphigus syphilitics, congenital pneumonia, atelectatic pneumonia, neonatal skin rash.

at 1: 2560. Hepatitis B and anti-HIV were non-reactive. At birth, the baby had severe respiratory distress and asphyxia. The baby presented with cyanosis, tachypnea, chest retractions, poor peripheral circulation, and oxygen saturation at 55%. Apgar scores were 3 and 5 at 1 and 5 minutes, respectively.

On examination, birth weight was 2.4 kilograms, length 45 centimeters, and head circumference 33 centimeters. Diffuse erythematous with desquamation, multiple fissures, and pale blistered lesions were prominent on palms and soles. (Figure 1, 2) No oral, ocular, nail or mucosal lesions were noted. The abdomen was slightly distended. Hepatosplenomegaly was found with the liver palpable 3.5 cm below the costal margin and the spleen tip palpable 2.5 cm below the left costal margin. Ronchi and decreased breath sounds were noted on thorax examination. Heart sound was regular, and a murmur was not detected. There was no lymphadenopathy and face dysmorphic. Other examinations were unremarkable.

The baby was intubated, but no significant improvement in oxygen saturation, and still had rapid breathing. Babygram, blood gas analysis, and complete blood count were taken. X-ray showed pneumonia with severe atelectasis of the left lung (Figure 3). Laboratory showed severe respiratory acidosis (pH: 6.8), leucocytosis (46.420/μL), thrombocytosis (414,000/μL), lymphocytosis (40.3%), monocytosis (16%), and TPHA reactive at the same level with mother (1: 2560). Antibiotic penicillin procaine at 50,000U/kg/dose intramuscular and broad-spectrum intravenous antibiotic, cefotaxime 100 mg/kg/day was prescribed. However, due to severe acidosis and collapsed lung, at 12-hours-age the baby did not survive.

**DISCUSSION**

Congenital syphilis is caused by *Treponema pallidum* infection through vertical transmission from mother to fetus. Risk factors of maternal syphilis include young age, low social economy, poor antenatal care, multiple sexual partners, have another sexually transmitted disease (STI), and narcotic usage. Transplacental
transmission is more frequent in mothers with primary or secondary syphilis than latent syphilis. Congenital syphilis often leads to spontaneous abortion, intrauterine growth retardation/small for gestation, non-immune hydrops fetalis, stillbirth, prematurity, and perinatal death.

Congenital syphilis is divided into early and late congenital syphilis. Early congenital syphilis has the onset of clinical manifestation before two years old, while the latter has an onset after two years of age. The majority, 70% of the infants with congenital syphilis, are asymptomatic at birth. Some are symptomatic with diverse presentations and multigorgan involvement. Mucocutaneous involvement is present in about 70% of infants with early congenital syphilis, and it is classically a vesiculobullous or maculopapular rash occurring on the palms and soles. Mucous patches in the perioral and perianal areas may also occur, and it is known as condylomata lata. Vesiculobullous lesion prominently affecting hands and feet, known as pemphigus syphilitics, and it is a rare presentation of congenital syphilis. Because the bullae are fragile and rapidly erode, denuded bullae and desquamation is the most likely clinical presentation, as in this case. The early rash is small blisters on the palms and soles, while a later rash is a copper-colored, flat, or bumpy rash on the face, palms, and soles. In this case, the characteristics were identical for the later rash of pemphigus syphilitics, and this lesion is considered highly contagious. Other reported lesion types of congenital syphilis include condylomata lata, annular, and erythema multiforme-like targetoid.

Other manifestations of early symptomatic congenital syphilis include irritability, poor feeding, snuffles/ syphilitic rhinitis, hepatitis, splenomegaly, anemia, jaundice, metaphyseal dystrophy, periodontitis/osteochondritis, nephritis/ nephrotic syndrome, and failure to thrive. Fatal case of congenital syphilis is often associated with congenital pneumonia, which is often complicated to sepsis and respiratory failure. Chest radiograph of congenital syphilis is often present as a course nodular pattern (2 – 5 mm in diameter) predominating in the basal regions of the lung. The pneumonitis caused by congenital syphilis is usually called pneumonitis alba. Twining and Kerley (1951) recognized four types of radiological picture: (1) interstitial fibrosis of linear type (2) gummatus appearing as a tumor (3) pneumatic or atelecatic, and is nearly always confined to infants with congenital syphilis (4) glandular, which is due to enlargement of the bronchial glands. As in this case, the patient present atelectatic of left lung and multiple coarse nodular (2-5 mm) typical for gummias of the right lung. Radiology features are consistent for congenital syphilis. In this case, severe pneumonia and collapsed lung has lead to sepsis and severe respiratory distress with lethal respiratory acidosis.

Hepatosplenomegaly is often found in congenital syphilis. Hematological evaluation of congenital syphilis usually manifests as anemia, leukocytosis, lymphocytosis, monocytosis, and thrombocytopenia. As in this case, neonate present with hepatomegaly without jaundice and splenomegaly, leukocytosis, lymphocytosis, monocytosis, but not anemia (Hb: 12 g/dL). The anemia of congenital syphilis has been attributed to a variety of causes, such as hemolysis, hypersplenism, or hypoplasia of bone marrow. Acute hemolytic anemia may occur early due to infection or sepsis. Splenomegaly is commonly found in congenital syphilis. An enlarged spleen may lead to hyperfunction and produce anemia. Aplastic type of anemia is reported in some cases due to generalized marrow hypoplasia. Aplastic type has persistent anemia, low reticulocyte count, and splenomegaly.

Serologic tests for syphilis are classified into non treponemal and treponemal tests. Non treponemal tests include the Venereal Disease Research Laboratory (VDLR) test and the rapid plasma regain (RPR) test. Non treponemal tests should be performed on the mother and infant so the comparison can be made. Serum quantitative of nontreponemal serologic titer that is fourfold higher than the mother’s titer, or (3) a positive darkfield test of body fluid(s). They should receive either aqueous crystalline penicillin G (50.000 U/kg intravenously every 12 hours for the first week of age, followed by every 8 hours beyond seven days of age) or aqueous procaine penicillin G (50.000 U/kg intramuscularly once daily) for ten days. If the availability of aqueous or procaine penicillin G is compromised, ceftriaxone for ten days can be considered with careful clinical and serologic follow-up. In this case, the TPHA of the patient was reactive at the same level as the mother (1:2560). VDLR test was not performed due to financial problems. Reactive TPHA could be from maternal antibodies. However, due to clear manifestations of congenital syphilis infection and the absence of previous maternal treatment, this case was highly probable for syphilis infection. The baby was treated with penicillin as a drug of choice for congenital syphilis. A single intramuscular injection of benzathine penicillin G (50.000 IU/kg) could be given to neonates who have a normal CIs. TPHA test detects antibodies (IgA, IgM, and IgG) to T. pallidum. It is highly sensitive (86%) and specific (94%) in diagnosing syphilis infection. But, since IgG from the mother is passively transferred to the newborn, a reactive serologic test in the neonate does not necessarily indicate the infant is infected because it can be due to maternal derived IgG. However, due to its high sensitivity and specificity compared to the nontreponemal test, it can be used to exclude congenital syphilis if the serologic tests are non-reactive in the mother and the infant.
physical examination with nontreponemal serologic titer less than fourfold from the maternal titer if the mother was not treated before or inadequately treated.

CONCLUSION
Congenital syphilis has high morbidity and mortality, so prompt diagnosis and treatment are required. Familiarity with clinical manifestation is vital to early diagnosis. Neonates born with hepatosplenomegaly, skin rash manifestation as vesiculobullous lesions (mostly eroded) known as pemphigus syphiliticus, broad desquamation of the skin, and/or congenital pneumonia typically atelectatic type is considered as some early manifestation of congenital syphilis. Treatment should be started promptly if clinicians find this manifestation in the newborn with syphilis infected mother in order to reduce the disease morbidity and mortality.

ETHICS IN PUBLICATION
The patient’s parents have given their consent for case publication and any physical examination documentation and photographs.

CONFLICT OF INTEREST
The author reports no conflicts of interest in this work.

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AUTHORS CONTRIBUTIONS
All authors equally contribute to the study from the case selection recent management to the case, evaluating the outcome until reporting the case study results through publication.

REFERENCES