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Hydrops Fetalis of unknown etiology in a 32 weeks pregnancy which was diagnosed as congenital neurosyphilis

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Introduction

Congenital syphilis occurs when the spirochete Treponema Pallidum is transmitted from a pregnant woman through the placenta. It can result in stillbirth, prematurity or other multiple clinical manifestations. The congenital condition usually occurs to women with no or scant prenatal care. Humans are the only natural host for Treponema Pallidum and so transmission usually occurs through the placenta or direct contact with lesions. A 19 y/o G1P0 32.2weeks pregnant woman arrived to triage complaining of premature contractions. Her prenatal care history was of no significance and her first and second trimester screening including level II ultrasound (US) were within normal limits (wnl). Non stress test was performed and category III tracing was noted. US scan was performed and the biophysical profile (BPP) was found to be 2/8. Amniotic fluid index was note to be 33cm. Ascites and pleural effusion was observed. Hydrops Fetalis was diagnosed. An emergency cesarean section (CS) was performed. Maternal rapid plasma reagent (RPR) returned as 1:64 and the syphilis diagnostic test FTA-ABS returned positive. The infant was initially intubated in the OR for the lack of respiratory efforts. Lumbar puncture was performed and spirochetes were noted to be present in the cerebrospinal fluid (CSF). The mother was treated with 2.4 million units of benzoin penicillin. Congenital syphilis is a devastating congenital disease since it results in a disseminated inflammatory response. The condition should be diagnosis promptly and treatment initiated as fast as possible. Even though treatment is very effective, it does not always abolish the clinical manifestations which already occurred. Some manifestations may persist for life and disrupt significantly the infants' quantity and quality of life.

Case Report

A 19 y/o G1P0 at 32.2weeks arrived to triage complaining of premature contractions. The patient reported that the contractions started several hours earlier and are 3/10 in intensity. The patients' history was insignificant for any illness or hospital admission. Prenatal care history included a normal first and second trimester screening in addition to a normal level II US. Upon performing a Non-stress test (NST), category III tracing was noted. Base line was noted to be 160bpm, with absent variability and repetitive late decelerations. The patient was found to be contracting every 5 minutes. US was performed for BPP and was found to be 2/8. No breathing, bodily or limb movements were visualized and the fetus did not exhibit tone. The AFI measured 33cm. Ascites and pleural effusion were noted. Maternal fetal medicine (MFM) specialist was consulted and delivery was recommended. The patient was admitted for emergency CS. The surgery was uncomplicated. The patient delivered an infant in breech presentation, APGARS 5, 6, 6 weighing 1940 grams. The infant did not have any respiratory efforts, appeared pale and diffusely edematous. The infant was intubated in the operating room and was taken to NICU. The placenta was removed and sent to pathology. The pathological report stated and early third trimester placenta with delayed villous maturation and poorly perfused distal villi by fetal blood. Initial fetal laboratory tests were significantly abnormal. WBC, Hb, Htc and Plt were noted to be 30K/UL, 6g/dl, 21% and 19K/UL, respectively. AST was 2828IU/L and ALT 438IU/L. Total bilirubin was 4.8mg/dl and direct bilirubin 1.9mg/dl. Central venous line was established

and 2 packed red blood cell units were transfused. Abdominal US was performed and hepatosplenomegaly was noticed. Cardiomegaly was noted on fetal echocardiography. In the morning of post-operative day 1, the maternal RPR returned as 1:64. The patients' prenatal chart stated non-reactive RPR. FTA-ABS confirmatory syphilis test was sent and found to be positive. Upon further questioning, the patient reported being sexually active with multiple partners during pregnancy. Infant CSF was obtained and was positive for Treponema Pallidum. Treatment with penicillin was initiated immediately for both infant and mother. The patient was counseled and treated with 2.4 million units of intra muscular benzoin penicillin. Since the initial RPR was negative and the one at 32.3weeks was positive at 1:64, early syphilis was diagnosed. The patient denies any genital lesions at any part of the pregnancy. At the post-partum period, the level II US was once more reviewed by MFM finding no abnormalities.

Discussion

Congenital syphilis is a rare condition which is not seen often around the western world. The incidence in the United States last year fluctuated between 8-12 cases per 100000 live births. Most of the cases are usually due to the lack of prenatal care or disease acquisition after first trimester screening tests [1]. The rate of congenital syphilis is increased in women with HIV but the contribution of maternal co infection is not completely understood. Transmission of Treponema Pallidum is either transplacetally, through maternal blood or through direct contact with infected lesions during birth [2]. Transmission rate is higher during primary and secondary disease than in latent disease and decreases to only 2% after 4 years of untreated disease. As the spirochete enters fetal circulation, it disseminates into almost every organ. Tissue destruction is through inflammatory reaction. It might result in an isolated laboratory or radiographic abnormality or in a fulminant disease affecting multiple organ systems [3]. Early syphilis is defined by onset of clinical manifestations before 2 years of age. Approximately 60%-90% of live-born neonates with congenital syphilis are asymptomatic at birth. Symptoms most commonly develop by 5 weeks postpartum [4]. Infants born with clinical symptoms are a rare finding and depend upon the timing of intrauterine infection and treatment [5]. Most common findings usually include: hepatosplenomegaly, jaundice, nasal discharge (snuffles), rash, generalized lymphadenopathy, nonimmune hydrops, fever, myocarditis, pneumonia, pseudo paralysis of parrot, sepsis due to other bacteria, ophthalmologic manifestations, gastrointestinal manifestations and nephrotic syndrome. Symptomatic CNS involvement is a rare entity among infants with congenital syphilis especially inside the uterus. Symptomatic CNS has two overlapping presentations: acute syphilitic leptomeningitis and chronic meningovascular syphilis [6]. Long bone abnormalities may be the sole manifestation of infants born to mothers with untreated syphilis. These occur in 60%-80% of cases and usually appear at birth or several weeks postpartum. Laboratory changes which are usually visualized in the case of congenital syphilis including anemia, thrombocytopenia and leukopenia or leukocytosis. All three laboratory manifestations were expressed in this case. Blood transfusion was required initially to correct the anemia present. Screening tests for neurosyphilis are not sensitive enough and include CSF RPR, WBC count and protein. Their specificity is adequate at 90%, 88%, and 78% respectively. Examination of the Treponema Pallidum DNA by polymerase chain reaction (PCR) proved to be the most definitive diagnostic method of neurosyphilis. Our case is a pure example of Treponema Pallidum spirochete which diffused from the fetal blood stream to the CSF and was able to be isolated with PCR [7]. Late congenital syphilis is defined by onset of clinical manifestations after 2 years of age. The differential diagnosis is wide and several other mimicking conditions must be excluded. Those include amongst others toxoplasmosis, rubella and cytomegalovirus infection. In addition, Herpes simplex, neonatal sepsis, immune and non-immune Hydrops Fetalis are also top priority. Clinical suspicion for congenital syphilis should be triggered in cases of unexplained Hydrops Fetalis, enlarged placenta, and failure to move an extremity, persistent rhinitis, jaundice, hepatomegaly, anemia, thrombocytopenia, generalized lymphadenopathy and persistent maculopapular rash. What triggered our suspicion and prompted immediate delivery of the fetus was the unexplained hydrops of a healthy previously normal pregnancy and the ominous tracing and US scan. Penicillin antibiotic is the treatment of choice. Treatment should be initiated immediately upon diagnosis, in order to prevent long term manifestations, especially in the case on neurosyphilis. The earlier in pregnancy, syphilis is acquired, the higher the chances for long term clinical manifestations if the diagnosis is missed. In our case, the regimen recommended is 10 days of aqueous penicillin 50,000units/kg intravenously every 12 hours. Additional option, in the form of procaine penicillin 50,000units/kg intramuscularly every day also exists. Maintenance of minimal inhibitory concentration (MIC) of 0.03units/mL of penicillin in the CSF for 7-10 days is required to achieve cure [8]. If penicillin treatment missed for a single day, the entire treatment needs to be repeated. Desensitization is recommended in case of penicillin allergy. Congenital syphilis is a rare disease which is not seen too often in the industrialized world. The rarity of the condition makes the diagnosis of congenital syphilis challenging. Level of suspicion should always be high especially if fetal compromise of an otherwise normal pregnancy could not be explained. Even though, the treatment is relatively simple, lack of correct diagnosis and immediate treatment can result in devastating consequences to the infant.

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