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Case Report

Congenital rubella syndrome: a case report

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ABSTRACT

Currently, rubella virus infections are very rare in many countries, and may not be recognized promptly. However, congenital rubella syndrome still appears, often in infants of mothers coming from countries with suboptimal vaccination programs. We describe a case of CRS in a full-term baby girl born to a foreign mother who documented perinatal antibodies against rubella. The baby had the classical abnormalities seen in infants with CRS include congenital cardiac and ocular anomalies. The diagnosis was confirmed by positive rubella serology in both infant and mother. Vaccination is the only known prophylactic measure for congenital rubella syndrome.

Keywords: Cataract, Congenital infection, Congenital heart disease, Congenital rubella, Sensorineural hearing loss

INTRODUCTION

Rubella virus produces a very mild but extremely contagious disease, sometimes referred to as German measles. Rubella typically begins with mild fever and lymphadenopathy followed by a brief appearance of a generalized erythematous, maculopapular rash. However, half of all rubella infections may be asymptomatic. Rubella virus infection has devastating consequences when infects fetuses, particularly during the first trimester, resulting in miscarriage, fetal death, or a constellation of birth defects known as congenital rubella syndrome (CRS).[1] CRS involves almost every organ of the body. It manifests with both antenatal and postnatal growth retardation; in addition to many malformations such as hearing loss, blindness, cardiac defects, microcephaly, and high risk of intellectual and behavioral problems.[2] At present, CRS can be difficult to diagnose given the unfamiliarity of current pediatric and family medicine practitioners with the disease and its overlapping clinical presentation with other congenital viral infections such as cytomegalovirus.[3]

CASE REPORT

A full-term female infant, delivered via an elective lower segment cesarean section due to breech presentation and polyhydramnios. The mother was a 33 years old multigravida, who had no history of antenatal fever, rash, medical illness, or drug intake. Antenatal ultrasound on the day of delivery revealed a single viable fetus with no obvious congenital anomalies. The baby was a product of non-consanguineous marriage and there was no family history of medical illnesses. At birth, the baby had respiratory distress, abdominal distension, and hemorrhagic skin eruptions all over the body. Apgar scores at 1 and 5 min were 7 and 8 respectively. The baby received routine neonatal resuscitation care and after stabilization, she was shifted to NICU for admission. Birth weight was 3.6 kg, head circumference was 36 cm, and the length was 52 cm (all at about 75th percentiles). Clinical examination revealed moderate-severe tachypnea with retractions and coarse crepitations. Heart sounds were normal with a grade III pansystolic murmur heard in the left infraclavicular and parasternal area. The abdomen

was moderately distended with palpable firm hepatosplenomegaly (liver 4 cm and spleen 3cm below the costal margins). There were multiple raised violaceous macules "blueberry muffin" spots, ecchymotic patches, and purpuric rashes all over the body (Figure 1). Examination of the eyes showed eyelids puffiness with bilateral subconjunctival hemorrhages, and both corneas were clear. There were no neurological abnormalities, no dysmorphic features, and no bleeding from any orifice. The rest of her physical examination was unremarkable. The baby received incubator care, oxygen via nasal prongs, and antibiotics (ampicillin and cefotaxime) after taking partial septic screening. She was kept NPO and was given IV fluids followed by total parenteral nutrition (TPN). The baby was placed on contact isolation because of suspected antenatal infections. She passed urine and normal colored meconium.

Figure1: The baby in her fifth day of life with widespread blueberry muffin spots, ecchymotic patches, and purpuric rash.

Figure 2: The baby in her 24th day of life with mild jaundice. All skin lesions disappeared.

The initial CBC showed WBCs 14.2x103 µL, hemoglobin 12.6 gm/dl, reticulocytes 10%, and platelet count $55\times10^3/\mu L$. Platelets dropped down to $22\times10^3/\mu L$, but later increased to normal levels. Other hematological indices maintained in average values. The blood glucose, electrolytes, and renal function tests were within the normal ranges. The liver function tests were moderately elevated. The urine test for reducing substances was negative. The coagulation profile was normal. Chest X-ray showed mild cardiomegaly with plethoric lung fields. TORCH serology and polymerase chain reaction (PCR) in blood were sent. Blood and urine cultures returned as negative. Brain ultrasound was unremarkable. Abdominal ultrasound showed moderate hepatosplenomegaly with no ascites. Echocardiography demonstrated the presence of apical ventricular septal defect (VSD), small patent ductus arteriosus (PDA), patent foramen ovale (PFO), and mild hypertrophy of right and left ventricles with good cardiac function. She developed jaundice within the first 24 hours, and she received phototherapy. A formal ophthalmologic evaluation revealed bilateral subconjunctival hemorrhages, semi-dilated pupils, clear right eye media, and normal retina. However, it confirmed the presence of cataract in the left eye. The neonatal metabolic screening was unremarkable. The TORCH screening results of the baby showed high positive rubella IgM and IgG antibodies. Unfortunately, the rubella virus polymerase chain reaction cannot be done. The rubella IgG serology of the mother was highly positive. The baby's serology for toxoplasma, herpes simplex, cytomegalovirus, and VDRL for syphilis was negative. Based on these findings the diagnosis of congenital rubella syndrome was made. On the 5th day of life the respiratory distress resolved, and the baby started on regular milk formula which was well tolerated. At the age of 10 days, the baby showed progressive conjugated hyperbilirubinemia (maximum total serum bilirubin was 319 µmol/L with a direct fraction of 227 µmol/L), and high liver enzymes (ALT 195 U/L, AST 419 U/L, ALP 228 U/L). The pediatric gastroenterologist advised starting lactose-free formula till getting the result of the galactosemia screening test. Repeated abdominal ultrasound revealed hepatosplenomegaly with a coarse liver echotexture, normal biliary system and gall bladder, and no ascites. The kidneys and the urinary tract were normal. GALT test for galactosemia in dry blood spot via enzymatic assay came as 10 GALT Units/gHb (normal reference >3.5 GALT Units/gHb). After the exclusion of galactosemia, we resumed regular milk formula. We started her on oral phenobarbitone; and later her bilirubin levels gradually regressed. Audiometry brainstem response testing was normal in both ears. The hemorrhagic eruptions gradually regressed and later disappeared. The baby progressively improved; and at the age of one month was discharged home in good condition. She was given appointments for follow up with a multidisciplinary team (neonatologist, cardiologist, gastroenterologist, audiologist, and ophthalmologist). At the age of 2 months, the baby was seen in the out-patient clinic, she was doing fine and her growth and development were appropriate for her age. Repeated echocardiography showed small apical VSD and small PDA with normal cardiac function. At the age of 3 months, the cataract in the left eye was more obvious (Figure 3). However, the baby traveled with her family back to their country to complete her treatment, and the

progress and outcome of her condition could not be assessed.

Figure 3: At the age of 3 months. The left eye cataract was more obvious.

DISCUSSION

Rubella virus is a single-stranded RNA virus belonging to the Togaviridae family and is the sole member of the Rubivirus genus. The virus is transmitted via airborne droplets and infected secretions. Humans are the only known host.[4] Effective vaccination programs as well as high-quality surveillance of rash/fever diseases have virtually eliminated congenital rubella syndrome in many countries. Rubella cases in developed countries are mainly "imported" from those countries where rubella is still endemic due to inadequate vaccination.[5] The mechanisms by which the rubella virus causes fetal damage are poorly understood. However, the rubella virus spreads through the placenta to the fetus then widely disseminated within the fetal tissues causing vasculitis and tissue necrosis. The direct viral damage is another mechanism, which occurs secondary to the reduced mitotic activity of the infected cells in the early fetal period. Rubella infection is progressive even after birth until the virus is eradicated from the body.[6] The outcome of fetal infection is dependent on the gestational age at the time of maternal infection, but fetal infection can occur at any stage of pregnancy. Infection occurring in the first 12 weeks of pregnancy causes congenital rubella infection in 90% of fetuses, with almost a 100% risk of congenital defects. From 13 to 17 weeks the risk of infection is about 60%, and the risk of defects is about 50%. From 18 to 24 weeks the risk of infection is about 25%, with hardly any risk of congenital defects.[7] The classic triad of congenital rubella syndrome is sensorineural deafness (58% of patients), eye abnormalities (43% of patients), and congenital heart diseases (50% of patients). Other manifestations include low birth weight, microcephaly, hepatosplenomegaly, bone marrow involvement, long bone radiolucencies, thrombocytopenic purpura, and the characteristic "Blueberry Muffin" spots, which represent dermal extramedullary hematopoiesis. There are also late complications such as diabetes mellitus, thyroid disorder, and subacute panencephalitis.[8] Congenital heart diseases may include PDA, the most common lesion, VSD, ASD, and pulmonary artery or valvular stenosis. Ocular abnormalities may include microphthalmos, congenital cataract, and the characteristic "salt and pepper" chorioretinitis that may present in 50% of cases.[9] The child is also predisposed to growth and developmental delay, learning disabilities, and a large number of psychiatric manifestations include autism and schizophrenia.[10] The diagnosis of congenital rubella syndrome can be confirmed by the detection of rubella-specific IgM in the cord blood or the serum collected within the first 6 months of life. It can also be confirmed by demonstrating persistent or increasing serum concentrations of rubella-specific IgG over the first 7 to 11 months of life. Detection of rubella virus RNA by reverse transcriptase-polymerase chain reaction (RT-PCR) in nasopharyngeal swab or urine provides laboratory evidence of CRS.[11] More recently, oral fluid testing for rubella-specific IgM or RT-PCR for rubella antigen has been very helpful in the diagnosis of CRS.[12] Active childhood immunization is not enough to prevent congenital rubella infection. However, vaccination of suspected childbearing women plays a paramount role in eliminating or reducing the incidence of this dreadful disease. Conception should be delayed for three months after rubella vaccination to avoid the risk of fetal infection.[13] There is no specific treatment for congenital rubella syndrome, and a multidisciplinary approach to improving the outcome and quality of life is usually adopted. Long-term follow up with a paediatrician, ophthalmologist, cardiologist, audiologist, and speech pathologist should be considered for early detection and management of late complications.[14]

CONCLUSION

Rubella is a vaccine preventable infection; and effective vaccination programs in many developed countries lead to much decline in the estimated number of CRS cases. In contrast, many cases of CRS are still found in developing countries where the rubella-containing vaccine has not been introduced in the national immunization programs or vaccination coverage is low. Some parents continue to believe that the MMR vaccine may cause autism, even though numerous data and studies have shown that there is no association. We reported this case to highlight the importance of vaccination as well as the high-quality surveillance of rubella, aiming at the eradication of rubella infection and congenital rubella syndrome globally.

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