Case Report

Congenital varicella syndrome in a neonate

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ABSTRACT

Congenital varicella syndrome is an extremely rare disorder occurring in <2% of the babies born to women infected with varicella between 7 and 28 weeks of pregnancy. The characteristic symptoms consist of skin lesions in a dermatomal distribution, neurological defects, eye diseases, and skeletal anomalies. We present a case of a newborn male baby who was shifted to neonatal intensive care unit.

Key words: *Congenital varicella syndrome, hypoplastic limb, neonate*

Congenital varicella syndrome is an extremely rare disorder occurring in <2% of the babies born to women infected with varicella between 7 and 28 weeks of pregnancy [1]. The risk of transmission to the fetus is approximately 2%, and it is more common when the mother has an infection in the 13th and 20th weeks of gestation [2]. The characteristic features include low birth weight, cutaneous scars in dermatomal distribution (76%), the absence of skin on a limb, hypoplasia of one or more limbs, malformed digits, and neurological defects (60%) including seizures, mental retardation cortical atrophy, hydrocephalus, cerebellar aplasia, encephalomyelitis, and dorsal radiculitis. The characteristic scarring may represent the cutaneous residue of the varicella zoster virus (VZV) infection of the sensory nerves analogous to herpes zoster. The virus may select tissues that are in a rapid developmental stage, for example, limb buds, which may result in one or more shortened and/or malformed extremities. Often, these atrophic areas are covered with a cicatrix. The lesion typically follows one or several nerve territories, suggesting that damage results from *in utero* zoster following primary fetal infection. The baby can have ocular abnormalities which include chorioretinitis, cataracts, microphthalmia, Horner's syndrome, and nystagmus. The musculoskeletal abnormalities are hypoplasia of the bones and muscles and/or absent or malformed digits. Genitourinary abnormalities occur in 12% and cardiovascular anomalies occur in 8% of babies.

CASE REPORT

Male newborn, born to non-consanguineous parents at 38 weeks of gestation with a birth weight of 2 kg and having normal Apgar scores, was shifted to neonatal intensive care unit in view of the hypoplastic left lower limb. On examination, the baby had cicatrizing skin lesions over the left lower limb and the gluteal region (Fig. 1). Infantogram done showed hypoplastic bones in the left lower limb with reduced soft tissue and hypoplastic left hemipelvis. Ophthalmic examination, two dimensional 2D ECHO and neurosonogram were normal. On inquiry, the mother had a history of chickenpox in the first trimester. Hence, in suspicion of fetal varicella syndrome (FVS), VZV-specific IgM and IgG were sent. Varicella IgM was negative and varicella IgG was raised (3079 mIU/ml) in the baby. The toxoplasmosis, rubella cytomegalovirus, herpes simplex, and HIV profile were negative, and the diagnosis of congenital varicella syndrome was made in view of characteristic history and clinical features.

DISCUSSION

Maternal varicella occurring between 7 and 28 weeks of pregnancy may lead to either spontaneous abortions or FVS. FVS has been reported under various names such as congenital varicella zoster syndrome, varicella embryopathy, and varicella embryofetopathy. The abnormalities in congenital varicella syndrome were reported to occur either directly due to cicatrizing lesions or secondary due to denervation of limbs. On an average, 30% of babies born with these lesions die during the 1st month of life, and there is a 15% risk of developing herpes zoster between the 2nd and 41st months of life [3].

Among women, who develop varicella during the first trimester, FVS can be diagnosed with the help of ultrasound and amniocentesis. Principal ultrasound signs in the prenatal diagnosis of FVS are ventriculomegaly, cerebral hypoplasia, microcephaly, calcifications, porencephaly, limb hypoplasia, intrauterine growth restriction, and polyhydramnios. Since the clinical features are due to reactivation of the VZV, time will be needed for this damage to manifest. Hence, ultrasound should be performed after 5 weeks from the time of maternal rash. Varicella zoster-specific polymerase chain reaction of the amniotic fluid aids in the diagnosis of congenital varicella syndrome [4,1].

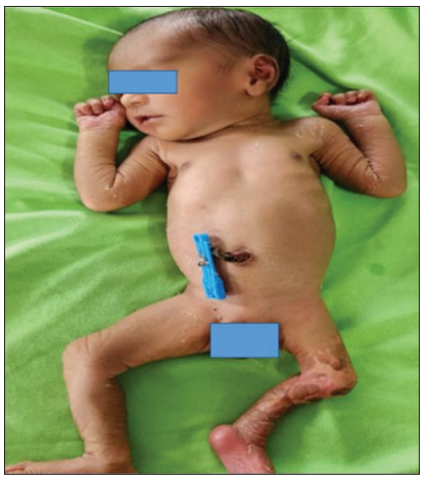

Figure 1: Hypoplastic left lower limb with cicatrising skin lesion

A major advantage of the invasive approach is to reassure over 90% of parents that their child has no risk of FVS or of any long-term impairment [3]. One disadvantage is the risk of fetal loss due to amniocentesis, although the risk is very low. Conversely, in the non-invasive approach, ultrasound lacks sensitivity and specificity as a diagnostic tool [5].

The criteria for the diagnosis include maternal varicella infection during pregnancy, the presence of congenital skin lesions that correspond to dermatomal distribution, and immunological evidence of *in utero* varicella zoster infection. The maternal infection will be considered positive by the demonstration of anti-VZV-specific IgM.

Since varicella infection, during the first trimester, is associated with serious malformations, attempts should be made to prevent varicella virus infection during pregnancy. All pregnant women should be enquired about immunity to varicella during their first antenatal visit. Moreover, those with negative history should be advised to avoid exposure to individuals with chicken pox. Pregnant women found to be non-immune should receive varicella zoster immunoglobulin after the exposure as early as possible ideally within 72–92 h [6,7].

Varicella vaccine is an attenuated live virus, which is safe and highly efficacious, preventing 90% of cases of varicella. VZV-related complications can be prevented not only in adolescent girls but also in their future offspring (s) by identifying and immunizing the girls who never had chickenpox. Being live attenuated virus, it is contraindicated in pregnancy, and hence, pregnancy should be avoided for 1 month after vaccination. When varicella occurs in pregnancy, there are indications to treat the mother with acyclovir for her own health. Although indications not clear, acyclovir has also been considered for post - exposure prophylaxis.. This strategy has the potential to prevent or attenuate VZV infection and would thus be an alternative to immune globulin for susceptible women exposed to varicella who did not receive immune globulin, due to delay or lack of availability, especially in late pregnancy [8,9].

CONCLUSIONS

FVS is rare but can have devastating consequences. The reason for reporting this case is to stress the importance of vaccinating all children with varicella vaccine as this disorder is associated with a high risk of morbidity and mortality. FVS is a preventable disease, and the best primary prevention of this disease is vaccination before pregnancy.

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