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Congenital Varicella Syndrome with Postnatal Reactivation of Varicella Zoster Virus

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Dear Editor,

Congenital varicella syndrome (CVS) occurs in approximately 1-5 per 10,000 pregnancies, if primary maternal varicella zoster infection occurs within the first twenty weeks[1]. CVS has approximately 30% mortality rate in the first few months of life and 15% risk of subsequent herpes zoster virus between the 2nd and 41st months of life[2].

We report a growth restricted male infant born at 32+5 weeks with microphthalmia. Primary maternal varicella infection occurred in the second trimester; this was not disclosed during the late antenatal booking at 25 weeks gestation. Respiratory distress was present at birth requiring ventilation. He deteriorated further on day of life (DOL) 9; developing neutropenic sepsis, pneumonitis and coagulopathy. Serum PCR studies were sent on DOL 13. Cranial ultrasound demonstrated diffuse increased echogenicity, loss of grey-white matter differentiation, bilateral periventricular cystic changes and ventriculomegaly. Abdominal ultrasound scan revealed multiple liver calcifications. Ophthalmology assessment revealed bilateral widespread chronic chorio-retinal scarring. Progressive hydrocephalus was evident radiologically and clinically. Phenobarbital was required for seizures.

A diagnosis of postnatal reactivation of congenital varicella was confirmed with positive PCR results in serum, cerebral spinal fluid and respiratory secretions for varicella zoster virus (VZV) DNA on DOL 15. Waning cell-mediated immunity to VZV in immunosuppressed individuals is thought to trigger the reactivation of herpes zoster virus[3]. Serum from the mother's late booking visit and previous pregnancy booking samples were retrospectively tested for VZV serology. This identified seroconversion from VZV IgG negative to positive, and a weakly positive varicella IgM result at the time of booking.

CVS is highly contagious via droplet and aerosols. Contact tracing was conducted due to risk to other infants in the neonatal intensive care unit. For identified contacts, the mothers' stored booking serology was tested for VZV IgG.

If the infant had risk factors for absence of trans-placental antibody (<28 weeks gestation, <1000grams, >60 days in NICU or received packed red cell transfusion), then serology for VZV was also performed for the infant. Four infants required varicella immunoglobin prophylaxis. No additional cases were identified.

Despite three weeks of intravenous aciclovir, VZV persisted in the CSF and the patient did not survive. A literature search of antenatal varicella screening in 5 countries (Ireland, United Kingdom, United States, Canada and Australia) highlights that routine screening is not universal. However, questioning maternal past history of VZV or vaccination is recommended for all women at their booking visit and if non-immune, counselling is vital regarding action if exposure occurs. The woman should immediately inform healthcare workers if potential exposure. Public health implementations have recommended that immunoglobulin testing could be offered to women without history of VZV. A safe, effective and routinely available vaccine could eliminate this life-threatening condition. The United States introduced the VZV vaccine in 1995 and studies report a marked decline in mortality[4].

This case highlights public awareness regarding the severity of VZV in un-immunized mothers and the importance of notifying a health professional if primary maternal chickenpox occurs in pregnancy. Moreover, testing for viral pathogens, should be considered in the septic workup for neonates where the cause is unknown and particularly where the response to antibacterial treatments is suboptimal.

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