Clinical Brief

Congenital Varicella Syndrome/ Vericella Zoster Virus (VZV) Fetopathy

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ABSTRACT

An 18 hour old female newborn born to a 3rd gravida HIV-ve mother, presented with a large erythematous patch of skin on right forehead and hazy right eye since birth.There was history of chicken pox in mother during fourteenth week of pregnancy. **[Indian J Pediatr 2010; 77 (1) : 92-93]** *E-mail: kkrhdang@gmail.com*

Key words: *VZV Fetopathy; Congenital varicella syndrome*

REPORT OF CASE

An 18 hour old female newborn presented with a large erythematous patch of skin on right forehead and hazy right eye since birth. Baby was born to a 3rd gravida HIV-ve mother by breech presentation vaginally without any other significant perinatal event. There was history of chicken pox in, mother during fourteenth week of pregnancy for which she had received only two doses of paracetamol. Examination revealed a term LBW, SGA baby weighing 1.75 kg. There was a 10 cm × 7 cm patch of raw skin with healthy granulomatous tissue over right scalp and forehead including the right eye. The right upper eyelid was deficient and without eyelashes. There was marked conjunctival congestion and cornea was completely opacified and vascularised. On the anterior aspect of right elbow there was a large (2cms diameter) partly ruptured bulla. The right lower limb was atrophic and shorter by l.5cm as compared to left. No other obvious congenital malformation was seen. Both skin lesions healed with cicatrisation over the next two weeks. Cornea healed with complete opacification.

Approximately 25% of the fetuses are infected when chickenpox is acquired during pregnancy. However only 2% of fetuses demonstrate VZV fetopathy when varicella occurs in the first 20 weeks of pregnancy.[1] The period of greatest risk to the fetus correlates with the gestational period when there is innervation and development of limb buds and eyes. Therefore its stigmata mainly involve the skin, extremities, eyes, and brain.[2] The characteristic cutaneous lesion known as cicatrix, is a zig zag scarring often in the dermatomal distribution.[1] The infant may also have varying degrees of aplasia of the brain. Involvement of eye may lead to cataract, chorioretinitis, corneal opacities, microphthalmia, and optic atrophy.[3] Viral damage to the sympathetic fibers in the cervical and lumbosacral cord may lead to Horner's syndrome, dysfunction of the urethral and anal sphincters respectively. The characteristic scarring may represent the cutaneous residua of the VZV infection of the sensory nerves analogous to herpes zoster.[4] The virus may select tissues that are in a rapid developmental stage *e.g.*, limb buds, which may result in one or more shortened and/ or malformed extremities. Often, these atrophic areas are covered with a cicatrix. The diagnosis of varicella zoster virus fetopathy is based mainly on the history of gestational chickenpox combined with stigmata seen in the fetus.[5] The damage caused by the fetal VZV infection does not progress in the postnatal period, which is an indication that there is no further viral replication or shedding. Many infants with severe manifestations of congenital varicella zoster fetopathy and aplasia of brain have significant neurological deficits, whereas those with isolated stigmata, amiable to treatment develop normally throughout childhood.[5]

VZV related complications can be prevented not only in adolescent girls but also in their future offsprings by identifying and immunizing the girls who have never had chickenpox. If a pregnant woman is not immune to varicella and has had a significant exposure, she should receive VZIG as soon as possible.

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Though VZIG prevents or attenuates the disease in the mother, it is not yet clear whether it can prevent fetal infection. Enders *et al* followed a cohort of 97 pregnant women with chickenpox who received VZIG and observed that none of their newborns had varicella fetopathy.[1] However, the number of enrolled women was not statistically significant to derive any conclusion. Further, there also exists an isolated case report of varicella zoster virus fetopathy in a newborn delivered by a mother who had received VZIG four days after exposure at 11 weeks of pregnancy and who herself manifested two weeks later with a mild varicella.[6] Hence any non immune expectant mother who has had a significant exposure to chickenpox at any gestation should receive VZIG , after she is informed that it may not alter the risk of congenital varicella syndrome.[7]

Management of chickenpox during pregnancy should include acyclovir when benefits to the mother outweigh the risks to fetus. American Academy of Pediatrics does not advocate oral acyclovir for pregnant women except in instances of serious viral mediated complications *e.g.*, pneumonia and encephalitis where intravenous acyclovir is recommended. The efficacy of acyclovir therapy of pregnant women for preventing or modifying the severity of varicella zoster fetopathy is not yet clear. Further, experimental data in animals have not proven the teratogenicity of acyclovir and controlled trials in pregnant women are not available , hence currently acyclovir is classified as Category C drug by US FDA for use in pregnancy.[8] Finally since the damage caused by fetal VZV infection does not progress in the post partum period ,antiviral treatment of infants with congenital varicella syndrome is not indicated.[5]

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