Congenital varicella–zoster virus infection

A rare case of severe brain and ocular malformations without limb or cutaneous involvement in a newborn after maternal subclinical infection

Yousef A. Al-Katawee, MD, Yousef A. Al-Hasoun, MD, Mohamed N. Taha, MD, Khalid Al-Moslem, MD.

ABSTRACT

Although congenital varicella-zoster virus (VZV) infection is rare, it carries serious morbidity and mortality to the fetus and newborn infant. We report a full term female newborn infant, born to a multipara unbooked mother who had VZV subclinical infection during the first trimester of pregnancy. Routine newborn examination showed cystic malformation of the left eye, and absence of the right eye globe. Radiological work up revealed severe brain and eye malformations, serological studies of both mother and baby were positive for VZV. The baby underwent palliative surgery to the eyes, upon discharge, a plan of multidisciplinary team was made for follow up including neurologist, ophthalmologist, pediatrician and social worker. Congenital VZV infection can be severe enough to cause catastrophic fetal anomalies and damage to the vital organs as many of those infants die in infancy.


Maternal infection with VZV early on during pregnancy (namely, up to 20 weeks gestation) can produce the characteristic malformations of the skin, the brain, the eyes, the extremities, or, rarely, other areas of the body. More over VZV subclinical infection of the pregnant woman can occur after close contact and exposure, which may lead to infection of the fetus. The maternal history, the clinical and the radiological findings as well as the results of laboratory studies were crucial in establishing the diagnosis of varicella-zoster embryopathy. The objectives for reporting this case are to highlight the importance of antenatal follow up and the need for more awareness and attention from the physicians and primary health care personnel towards the pregnant women particularly at first and second trimester of pregnancy to avoid VZV infection to the fetus, and its subsequent serious sequela to the newborn infant.

Case Report. A 3.15 kg, full-term baby girl was born by spontaneous vaginal delivery, apgar scores were 9 and 10 at the first and fifth minutes to a 42-year-old unbooked grand multipara mother (P7+0). The pregnancy was uneventful, no history

From the Department of Neonatology, Al-Yamamah Hospital, Riyadh, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Yousef A, Al-Katawee, Consultant in Pediatrics & Neonatology, Department of Neonatology Al-Yamamah Hospital, PO Box 106208, Riyadh 11666, Kingdom of Saudi Arabia. Tel. +966 (1) 4914444 Ext. 1325/1326, Fax. +966 (1) 4914896 E-mail: yrshmh@hotmail.com
of drugs to mother and there was no history of maternal illness apart from exposure to chicken pox during first trimester where the whole family, including the father, were affected except the mother who was asymptomatic, maternal previous history of chicken pox was uncertain and parents are non-consanguineous. Through routine newborn examination after birth (Figure 1), there was big soft cystic swelling measuring 4 x 5 cm located at the left eye. The right eye was not palpable. Head circumference was 35 cm and length was 49 cm. Other systemic examination including musculoskeletal, extremities and genitalia were normal. Frequent tonic seizures were observed on the 3rd day which was controlled on phenobarbitone.

Initial routine laboratory results as well as ultrasound of abdomen were normal. An MRI of orbits and brain (Figure 2) showed huge lobulated soft tissue and cystic density mass lesion occupying the left orbital cavity protruding to the anterior and direct continuation with the left temporal fossa through a widened left inferior orbital fissure. There was bilateral microphthalmia as well as optic atrophy. The brain study revealed a large cyst with hypoplasia of the cerebellum, agenesis of the corpus callosum and single widened central ventricular cavity (holoprosencephaly). Echocardiographic examination was carried out and showed a small secundum atrial septal defect. The infant underwent palliative surgery where the left eye intraorbital cyst was excised and small sized conformer was inserted in the right eye (Figure 3), postoperative findings confirmed the presence of severe bilateral microphthalmia. Histopathological and microscopic examination of the cyst architecture revealed irregular fibro-vascular tissue in some areas and primitive retina-like structures as well as myxoid tissue in other areas, fluid was identified as cerebrospinal fluid. Maternal and infant’s serological studies as well as infant’s cerebrospinal fluid were positive for varicella-zoster specific IgG antibodies, specific IgM antibodies were negative. The infant’s postoperative period was uneventful, and she had a satisfactory general condition upon discharge. At present, she is attending outpatient clinic visits on a regular basis.

Discussion. Approximately 1-2% are the risks of fetal malformations caused by maternal varicella infection during the first 20 weeks of pregnancy.\textsuperscript{1,2} Maternal varicella subclinical infection and congenital varicella syndrome have been reported.\textsuperscript{3} The mother in our report was considered to have subclinical infection as there was clear history of exposure to the virus in her first trimester of pregnancy where all members of her family including her husband had chicken pox at this
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been used, but are neither specific nor sensitive for differentiation of congenital varicella syndrome from benign congenital chicken pox.\textsuperscript{12} The damage caused by fetal VZV infection does not progress post partum, an indication that there is no persistent viral replication so infants with varicella embryopathy do not require isolation and antiviral treatment is not indicated.\textsuperscript{4} Acyclovir treatment may be given to the mother with severe varicella, however neither it’s safety nor it’s efficacy for the fetus is known.

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