Case Reports

Congenital varicella syndrome

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ABSTRACT

We report a case of a premature baby, 35 weeks gestation. Whose mother presented with skin rash during pregnancy. The newborn patient presented with cutis aplasia, rudimentary digits in both feet with pansystolic murmur, microphthalmia and normal fundoscopic examination. Chest x-ray showed plethoric lung fields and echocardiogram confirmed a ventricular septal defect. The findings are consistent with congenital varicella syndrome.

Keywords: Congenital varicella syndrome.

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Varicella zoster fetopathy is a very rare syndrome occurring in no more than 2% of infants born to mothers who had varicella infection during the first and second trimesters of pregnancy. Diagnosis of varicella fetopathy is based on history of maternal chicken pox infection during pregnancy and the typical stigmata of congenital varicella in the baby both of which are well documented in the patient that we are describing. As there is no progression of the virus following delivery, antiviral treatment is not indicated for infants with congenital varicella syndrome which, with the recent availability of the varicella vaccine should be preventable by immunization of Varicella Zoster Virus-susceptible young women in the child bearing age.

Case Report. We are describing a male neonate product of a 35-week gestational age, normal vaginal delivery, cephalic presentation who was delivered to a 33 year old Jordanian gravid 8, para 6 unbooked mother. The mother gave history of chicken pox during her pregnancy manifested by the typical skin rash that appeared at about 14 weeks of gestation for which she was seen by a general practitioner, she was reassured and no action was taken. The baby had an Apgar score of 8/10 at one minute and 10/10 at 5 minutes with no resuscitation required whatsoever. Physical examination following delivery showed a small premature baby with birth weight of 1700 gm, head circumference 31 cm, length 45 cm. Cutis Aplasia (Figure 1) and rudimentary digits involving both feet (Figure 2) were noted. Cardiac examination revealed a pansystolic murmur at left lower sternal border grade 3/6. Ophthalmological consultation documented the presence of microphthalmia (small cornea in an otherwise normal eye) and normal fundoscopic examination. Apart from the mentioned data the rest of the physical examination was normal including full neurological assessment.

Investigations conducted. Complete blood count showed white blood cell count of 8.800, mean corpuscular volume of 96.6fl, Hemoglobin of 13.6 g/dl, mean corpuscular hemoglobin of 32.7pg, packed cell volume of 40.2%, mean corpuscular hemoglobin count of 33.8g/dl and platelets of 150,000. Liver function test and electrolytes were normal. Skeletal survey showed absent phalangeal bones of both feet digits. Chest x-ray showed plethoric lung fields. Echocardiogram confirmed the presence of ventricular septal defect. Chromosomal studies showed normal male pattern 46 XY. Computerized tomography of the brain was normal.

Discussion. Transplacentual passage of the varicella virus can occur at any time during pregnancy. However congenital varicella syndrome is a very rare complication of maternal varicella
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Figure 1 - Showing cutis aplasia in the scalp of our patient, with its black center and surrounding hair loss.

Figure 2 - Showing rudimentary digits more prominent on the toes of the right foot.

infection occurring during the first or second trimester of pregnancy with a maximum fetal risk of 2% when maternal infection occurs between 13-20 weeks gestation and a risk of 0.4% when maternal infection occurs before 13 weeks gestation. Infection towards the end of pregnancy, especially a few days prior to delivery, results in clinical manifestations of disseminated varicella infection in about 25% of neonates. Sequelae of congenital varicella syndrome mainly involves the skin, extremities, eyes and brain. Manifestations include intrauterine growth retardation, intracerebral calcifications, hydrocephaly and microcephaly (central nervous system infection and damage). Cicatricial skin lesions and scars in dermatomal distribution (infection and damage to sensory nerves). Cataract, chorioretinitis, microphthalmia and optic atrophy (damage to optic stalk). Another hallmark is one or more shortened and malformed extremities of varying severity ranging from an entire limb hypoplasia to minor phalangeal deformity (cervical or lumbosacral cord damage).

Children with congenital varicella syndrome may also show features of sympathetic fiber damage including Horner syndrome, anal dysfunction and neurogenic bladder with concomitant vesicoureteral reflux and all its related complications. Congenital varicella syndrome is diagnosed on the basis of a positive history of maternal infection during pregnancy manifested by the typical skin rash of chicken pox and stigmata of varicella in the baby, both of these features are well documented in the patient we are describing. The virus cannot be cultured from the affected newborn, however viral DNA can be detected by hybridization and polymerase chain reaction II technique in formalin fixed tissue samples from lung, spleen and adrenal glands. Virus specific IgM can sometimes be detected in cord blood but it rapidly declines post partum, it is however used for antenatal diagnosis by obtaining and testing a fetal blood sample for varicella zoster virus-specific IgM titer. It is true that many infants with congenital varicella have severe neurological deficiencies however another group of patients may have only isolated stigmata. This explains the absence of CNS manifestations and cicatricial skin lesions in our patient who had the typical limb deformity manifested by rudimentary digits of both feet as well as microphthalmia documented by ophthalmological consultation. Cutis aplasia, although not a feature of congenital varicella syndrome, has been described in conjunction with this syndrome by other authors.

We believe that the positive maternal history of chicken pox during pregnancy combined with the stigmata of varicella fetopathy described in our patient are enough for describing and diagnosing this patient as congenital varicella Zoster virus infection fetopathy, a syndrome that with the recent introduction of varicella vaccine should well be prevented by immunizing young susceptible women in the child bearing age.

References