



Case Report

Congenital Varicella Syndrome with Congenital Heart Disease - A Rare Association

Ashoka C¹, Usharani S², Avinash T³, Kumar G. V¹, Viswanathakumar H.M⁴

¹Assistant Professor of Paediatrics, Sri Siddhartha Medical College, Tumkur, Karnataka.

²Assistant Professor of Community Medicine, Sri Siddhartha Medical College, Tumkur, Karnataka,

³Senior Resident in Hemato-Oncology, Kidwai Institute of Oncology, Bangalore, Karnataka

⁴Professor & HOD of paediatrics, Sri Siddhartha Medical College, Tumkur, Karnataka.

Corresponding Author: Ashoka C

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ABSTRACT

Congenital varicella syndrome refers to the septum of fetal anomalies associated with maternal varicella zoster virus (VZV) infection during the first trimester of pregnancy. We report a case of congenital varicella syndrome with congenital heart disease - a rare association.

Key words: Congenital varicella syndrome; congenital heart disease.

INTRODUCTION

Congenital varicella syndrome refers to the spectrum of fetal anomalies associated with maternal varicella zoster virus (VZV) infection during the first trimester of pregnancy. Laforet and Lynch first described this syndrome in 1947. [1] This syndrome is rare and risk to the fetus uncertain. We describe an unusual case of congenital varicella syndrome (CVS) associated with congenital heart disease (CHD).

CASE REPORT

A term small for date female baby was born to a 21 year old primigravida mother by caesarean section for breech presentation; with a birth weight of 2000 gm. Mother had chickenpox at the 12th week of gestation and was treated

symptomatically. Three antenatal sonograms done at the 14th, 18th and 32nd week of gestation did not pick up any anomaly, except for intrauterine growth restriction (IUGR). Baby had microcephaly and left microphthalmia with absent eye lashes. Cicatricial skin lesions were present over left frontal and maxillary regions (Trigeminal Nerve distribution) (Figure 1) and right sacral region. The baby had hypoplastic left upper limb and right lower limb (Figure 2). Neurological examination revealed normal fundus with hypotonia of involved limbs with absent deep tendon reflexes. Cardiac examination revealed harsh ejection systolic murmur best heard over left upper sternal border and left infraclavicular areas. Investigations showed Hb - 12.88 g%, Total leucocyte count of 22,100 cells/ cumm with 81% Polymorphs, 16 % Lymphocytes, 02 %

Monocytes and 01% Eosinophils. Echo showed large PDA (3.5 mm) with left - right shunt. Plain and contrast CT brain were normal. Diagnosis of congenital varicella



Fig : 1 Cicatricial skin lesion over left frontal and maxillary region.

syndrome was made based on history suggestive of acute varicella infection in the mother during first trimester, characteristic skin lesions and limb anomalies in the baby.



Fig : 2 Hypoplastic left upper limb and right lower limb.

DISCUSSION

Congenital varicella syndrome (CVS) is a rare disorder resulting from maternal - fetal VZV transmission, usually between 8 and 20 weeks of gestation. The incidence of maternal VZV infection has been estimated to be between 5 to 7 per 10,000 pregnancies. [2] Fetus specific factors play a crucial role and fetal consequences of maternal exposure may be highly variable. 2 % of fetuses infected between 8 and 20 weeks gestation may exhibit pathogenetic effect of VZV. [3] The CVS incidence rate to be about 2.8 - 4 cases per 10,000 pregnancies. [4] The clinical consequences in the offspring of a woman with chickenpox complicating pregnancy may take any of 3 distinct patterns (Table: 1) depending on the stage of pregnancy at which infection occurs. [5]

Table: 1. Chickenpox complicating pregnancy: foetal outcome

Period of maternal infection	outcome
1 st or 2 nd trimester	Congenital Varicella Syndrome
2 nd or 3 rd trimester	Childhood Herpes Zoster
Perinatal Period	Neonatal Chickenpox

VZV is well known for its neurotropic properties. The virus has also

been known to induce chromosomal abnormalities both in vitro and in vivo. [6] The precise mechanism of infection with VZV in utero is unknown. The spectrum of clinical manifestations of the syndrome has been fairly well delineated since the first report by Laforest and Lynch in 1947. Subsequently more such cases have been reported and the frequency of various clinical manifestations has also been well established (Table 2).

Table 2: Clinical manifestation of CVS. [7,8]

Characteristic features	Remarks
Small weight for date	
Cicatricial skin lesions	Zigzag scarring on dermatomal pattern*
Ocular manifestation	Microphthalmia*, cataract, optic atrophy, nystagmus, anisocoria, chorioretinitis.
Hypoplastic limb	Hypoplastic upper limb and lower limb*, hypoplasia or absence of digits, club foot.
Neurological manifestation	Microcephalus*, Cortical atrophy, mental retardation, abnormal EEG, Horner's syndrome, Facial paralysis.
Genito urinary anomalies	Hydronephrosis, hydroureter.
Gastrointestinal anomalies	Gastro-esophageal reflux, hepatic calcification, duodenal stenosis, immature left colon, Atresia of sigmoid colon, Intestinal obstruction.

* Features present in this baby.

This baby had many of the classical features of CVS such as IUGR, cicatricial skin lesions, ocular manifestations like microphthalmia and loss of eye lashes, limb anomalies like hypoplastic right upper limb and left lower limb with thin and elongated fingers. However the case we have presented is unique so far as the association of congenital varicella syndrome with congenital heart disease. A review of literature showed that association of congenital varicella syndrome and congenital heart disease has not been reported previously. Diagnosis of the syndrome is essentially clinical, based on history of chickenpox in the mother and recognition of the characteristic defects in the neonate.

CONCLUSION

This case is being reported for its rarity and unique association with congenital heart disease.

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