

Congenital Varicella Syndrome: Presenting with Eye Complications

Primary varicella infection during pregnancy can rarely result in intrauterine infection of the fetus, presenting as congenital varicella syndrome. We report the clinical features and complications of congenital varicella syndrome in a 40-day-old boy who presented with right corneal opacity and scalp lesion since birth, and diarrhea, fever and lethargy for 2 days.

The baby was born at term gestation, by a normal vaginal delivery, and had a birth weight of 2.5 kg. A cicatricial non-progressive rash with alopecia, involving the right side of scalp and forehead, was noticed since birth. On examination, a 7 cm erythematous plaque with central ulceration and white margin was seen on the right scalp and forehead (*Fig. 1*). Exophthalmos of the right eye with corneal opacity and lower motor neuron paralysis of right facial nerve were also present.

There was maternal history of papulovesicular itchy rash, which began on the trunk and soon became generalized during late first trimester, and lasted for 7-8 days. There was no history of contact or past history of similar rash.

Investigations of the patient showed a hemoglobin level of 9.4 g/dL, total leukocyte count of 15,700/cu mm, with 60% polymorphs. Cerebrospinal fluid examination showed clear fluid with 75 cells (80% polymorphs) and normal glucose and globulin levels. Blood and cerebrospinal fluid cultures were sterile.

Contrast enhanced cranial and orbital CT showed right cerebral porencephalic cyst, right cerebellar hypoplasia, flattening of posterior

pole of the right orbital globe with thinning of the optic nerve and atrophy of extraocular muscles. The CT features were suggestive of sequelae of an infarct involving right internal carotid artery (*Fig. 2*). The patient's specific IgG antibody (by ELISA) against varicella, obtained on 2 occasions two months apart showed rising titres. Specific IgM varicella antibody was negative (<8 U/mL).

A diagnosis of congenital varicella syndrome was made based on, history suggestive of acute varicella infection in the mother during the first trimester, characteristic skin rash and eye lesions, rising titer of varicella antibody and characteristic CT findings. The child received intravenous fluids for correction of dehydration and antibiotics for meningitis. Clinical improvement was noticed during the next 48 hours. Tarsorrhaphy was done to protect the eye.



Fig. 1. Cicatricial scalp rash and exophthalmos right eye with corneal opacity.

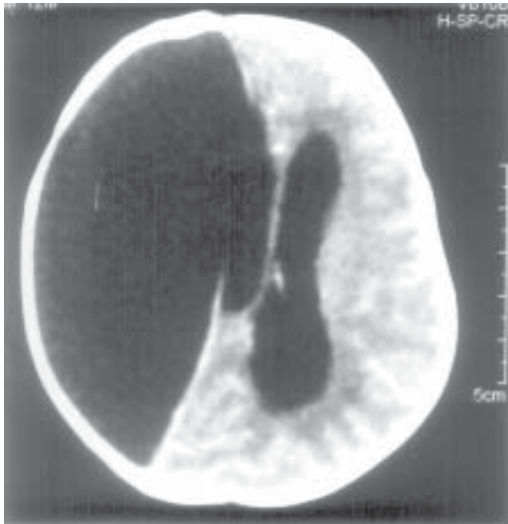


Fig. 2. Porencephalic cyst seen replacing right cerebral hemisphere on cranial CT.

Intrauterine infection with VZV within 20 weeks of gestation, resulting in congenital abnormalities, is rare with less than 100 cases reported in the literature(1). A small proportion (5-7%) of women in the reproductive age group is negative for varicella antibody(2). The risk of congenital varicella syndrome is between 0.4-2.0% when fetus is infected before 20 weeks' gestation(3).

The key event in pathogenesis of congenital varicella syndrome is the reactivation of virus in fetus, subsequent to primary infection, leading to zoster, which is responsible for clinical manifestations(4). Skin is the organ involved most frequently, and manifestations are found in three fourths of all affected patients. Nervous system and eye are involved in approximately two thirds and half of the patients respectively(1).

The present case highlights the debilitating effects of varicella during pregnancy. Administration of the live attenuated vaccine is very effective in reducing the risk of varicella and attendant morbidity. The Indian Academy of Pediatrics recommends that, wherever possible, the vaccine be administered to susceptible adolescents(5).

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