

Case Report

A Case Report & Management of Bilateral Congenital Cataract due to Congenital Rubella Syndrome

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ABSTRACT:

Congenital rubella syndrome (CRS) is referred to as the consequences of rubella infection *in-utero*. In India, it is assessed that about 50,000 children are diagnosed with congenital cataract every year. 25% cause of which is maternal rubella ¹ In first trimester if mother gets contacted with the rubella virus it results in abortion, stillbirth or an infant born with CRS. Cataract is the commonest finding seen in eyes. Others included bilateral or unilateral microphthalmos, iris hypoplasia, cloudy cornea, rubella retinopathy, nystagmus, concomitant strabismus, primary optic atrophy, and bilateral dacryostenosis. Authors here would like to report a case of 4 month old baby with congenital cataract which was diagnosed with congenital rubella syndrome, baby was operated for lensectomy with anterior vitrectomy, was kept aphakic and was given spectacles to prevent amblyopia.

Key words: congenital cataract, congenital rubella syndrome, rubella.

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INTRODUCTION

Congenital rubella syndrome (CRS), caused by rubella virus infection during pregnancy, remains a public health concern in developing countries. Three to five per cent of all suspected congenital cataract cases in India have been proven due to rubella infection. Only about 45%–60% of pregnant women and infants in India receive the rubella vaccination.

Author would like present a case of a 4 month old female who tested positive for the rubella immunoglobulin, she had no symptoms at birth. On ophthalmic examination, she was diagnosed with bilateral cataract. She was detected to have patent ductus arteriosus (PDA) on echocardiography.

CASE REPORT

A 4-month-old infant came to our ophthalmology OPD with reference from primary health care. Parents came with complaint of white eye reflex in eyes which noticed by them. A detailed history was taken.

Female infant born on 37 weeks' gestational age (based on mother's last menstrual period). She was born to a 27-year-old primigravida woman from Kolhapur Maharashtra. She initiated prenatal care at primary health care her village at approximately 20 weeks' gestation, where a fetal ultrasound (US) and serologies reportedly were normal. Mother did not have any history of febrile illness with associated malaise, upper respiratory symptoms and a widespread pruritic, vesicular rash. Mother had a normal vaginal delivery assisted by midwife. Father gives history of uneventful intra natal period. New born baby was then started with breastfeeding after half an hour of delivery. Mother after 1 week of birth of child noticed white reflex in both eyes for which she went to primary health care and she was referred to specialist's opinion. Ophthalmological examination of child was done under general anaesthesia which showed both eye cataract with normal anterior segment. Child was not following torch light nor

recognizing mother showing visual deprivation. Pupils of the child were then dilated with diluted mydriatics which was prepared. (3ml of 0.8% w/v tropicamide with 5% w/v phenylephrine with 7ml of 0.5% w/v carboxymethyl cellulose) for detailed cataract and retinal evaluation which showed central nuclear cataract and retina was seen through haze and appeared normal. USG B-scan was done showed no pathology in posterior segment. After which we have planned for bilateral lensectomy for prevention of visual deprivation amblyopia

Child was referred to paediatrician for detailed systemic evaluation including cardio-pulmonary status. General examination revealed child slightly irritable, physically underdeveloped with weight of 4.6kgs and no head holding for his age. Bounding peripheral pulse with pulse rate of 152 breaths/min. Breathlessness was present, with breath rate of 52 breaths/min. No cyanosis, icterus, clubbing was observed. Blood pressure was 100/50mm hg. Child was immunized till date. On cardiac evaluation, paediatrician observed heaving apex, systolic thrill below left clavicle, a machinery continuous murmur which was accentuated in systole hinting us towards Patent ductus arteriosus. Rest of the systems were within normal limits. Looking at the complete ophthalmological and paediatric evaluation we suspected congenital rubella syndrome.

INVESTIGATION

Complete blood count revealed anaemia with haemoglobin 8.7g/dl. Urine routine and microscopy, random blood sugar, serum urea and serum creatinine were normal. HIV and HbsAg test were negative. Liver function test were normal. Chest X-ray was normal and ECG showed arrhythmia. TORCH evaluation of the child was also carried out which was positive for Rubella with positive IgM

(1.28IU/ml)(normal < 0.8IU/ml) & IgG (160IU/ml) (normal <10IU/ml). 2D-Echo confirmed small 2mm patent ductus arteriosus with left to right shunt, with ejection fraction 60%.

DIAGNOSIS

The above clinical features along with positive investigation lead us to diagnose this baby with Congenital Rubella Syndrome (CRS).



Fig 1: Shows nuclear cataract in left eye.

MANAGEMENT

After diagnosing CRS, we went ahead with our plan towards lensectomy for which multispecialty consultation from paediatrician, cardiologist and anaesthesiologist were taken after which one unit PCV was given to improve the haemoglobin to 11.1g/dl. Cardiologist gave us the clearance for the lensectomy procedure. Baby was taken under general anaesthesia and lensectomy with anterior vitrectomy was performed in both eyes a week apart. Retinoscopy was performed after the second eye lensectomy under general anaesthesia. Spectacles with +16 diopteric power in both eye, were prescribed on 2nd post operative day. Postoperatively moxifloxacin and prednisolone combination eye drops were given in tapering dosage over the period of follow-ups.

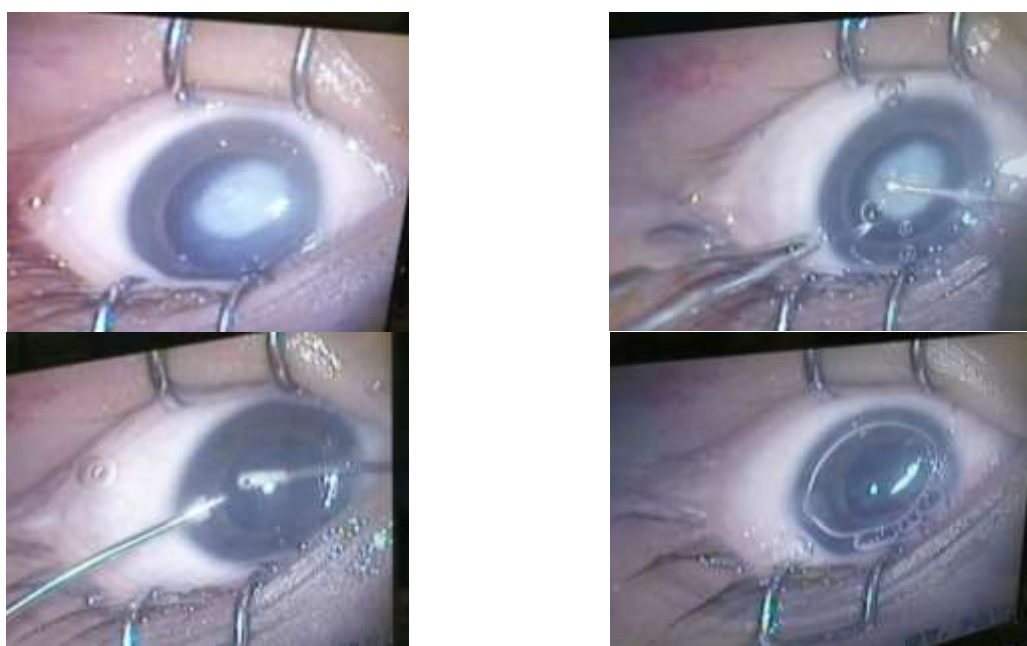


Fig 2: Intraoperative lensectomy with anterior vitrectomy.



Fig 3: Post-operative day 1 showing aphakia



Fig 4: Postoperative with spectacle correction.

DISCUSSION

Congenital rubella syndrome (CRS) is referred to as the consequences of rubella infection *in-utero*. In India, it is assessed that about 50,000 children are diagnosed with congenital cataract every year. 25% cause of which is maternal rubella¹ In first trimester if mother gets contacted with the rubella virus it results in abortion, stillbirth or an infant born with CRS. In first two trimesters, the foetus is susceptible to this infection due to the rapid organogenesis and undeveloped specific immunity against the virus. The severity depends upon the virulence of the organism and the timing of the foetal infection. The earlier in pregnancy the infection occurs, the greater is the damage to the foetus.² Incidence of birth defects seen when infection occurs within the first 10 weeks of pregnancy is reported to be 90%.^{2,3}

Table 1: Diagnostic criteria given by WHO for clinically confirming the CRS case is : Any infant in whom a qualified physician detects at least 2 of the complications listed in (A) OR 1 in group (A) and 1 in group (B)

GROUP A	GROUP B
Loss of hearing	Purpura
Congenital heart disease	Splenomegaly
Pigmentary retinopathy	Microcephaly
Cataract	Developmental delay
Congenital glaucoma	Meningoencephalitis
	Radiolucent bone disease
	Jaundice with onset within 24 hours after birth

In CRS almost all ocular structures are affected either in combination or individually. Rubella cataract is the most common ocular sign. The virus enters the lens before the development of the lens capsule that would otherwise act as barrier to the virus. Therefore, rubella cataract is always at the foetal nuclear level and is frequently bilateral.^{4,5} Congenital glaucoma is an uncommon finding following maternal rubella.^{6,7} Glaucoma may be caused either by failure of absorption of the mesoderm of the angle or by failure of the canal of Schlemm to differentiate.⁸ Incidence of retinopathy reported in CRS is 13.3% to 61%. The pigment deposits may vary from fine powdery, sprinkled or granular shapes throughout the retina, especially the posterior pole, or discrete patchy black lesions varying in size and location resembling retinitis pigmentosa. Foetal infection with rubella virus causes serious multisystemic malformations, resulting in severe morbidity and mortality. Cardiovascular system, central nervous system and sensori-neural hearing loss has been reported. Incidence of cardiac defects in CRS documented is 95%.⁶ Cardiac anomalies observed are patent ductus arteriosus, stenosis atrial septal defect, pulmonary stenosis and ventricular septal defect. CRS due to its wide range of ophthalmic and systemic disorders, poses a continuous challenge. By early diagnosis and intervention, the quality of life can be improved for these infants. Ophthalmologists can play a major role in the early diagnosis of this multisystem disease because the ocular findings can be detected at birth unlike most of the systemic manifestations such as hearing loss and neurological abnormalities. Proper immunisation nearly always prevents the disease.

Babies with congenital rubella syndrome who develop respiratory distress may require supportive treatment in the ICU.

Hepatosplenomegaly is monitored clinically. No intervention is required. Patients with hyperbilirubinemia may require phototherapy or exchange transfusions if jaundice is severe to prevent kernicterus.

True haemorrhagic difficulties have not been a major problem; however, IVIG may be considered in infants who develop severe thrombocytopenia. Corticosteroids are not indicated.

Infants who have a rubella-related heart abnormality should be carefully observed for signs of congestive heart failure. Echocardiography may be essential for diagnosis of heart defects. Contact isolation is required for patients with congenital rubella during hospitalizations because babies are infected at birth and are usually contagious until older than 1 year unless viral cultures have produced negative results.

Postnatal rubella treatment is supportive. No specific antiviral agent for rubella is currently available. Starch baths and antihistamines may be useful for adult patients with uncomplicated rubella and troublesome itching. For complicated cases, treatment is for severe arthritis affecting weight-bearing joints,

encourage rest. Nonsteroidal anti-inflammatory drugs (NSAIDs) may be helpful, but corticosteroids are not indicated. For patients with encephalitis, provide supportive care with adequate fluid and electrolyte maintenance. Thrombocytopenia is usually self-limited but, if severe, consider intravenous immunoglobulin (IVIG). Corticosteroids have not demonstrated any specific benefit. Splenectomy is not indicated.

A neurologic evaluation and follow-up care are needed for children who have CNS anomalies, including motor weakness and delay, poor balance, mental retardation, behavioural abnormalities, and learning deficits.

Adequate rehabilitation programs comprising physical and occupational therapy may be beneficial for patients with motor weakness and motor delay.

The combined effort of ophthalmologists and other health care personnel working in this field is required to achieve optimal results. A greater awareness of various aspects of CRS in our country is the need of the hour.

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