

Congenital Rubella Syndrome: It Still Exists in India

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ABSTRACT

We report here a case of congenital rubella syndrome in a child who presented with hearing loss, patent ductus arteriosus and coloboma of the eye. The child was evaluated and rehabilitated

using high gain hearing aid. This case is reported to highlight the persistence of infections like rubella which can be eradicated by vaccination and thus are avoidable causes of congenital hearing loss.

Key Words: Congenital rubella syndrome, Congenital hearing loss, Intrauterine infection

INTRODUCTION

Maternal infection with rubella in the first trimester of pregnancy which results in congenital rubella syndrome (CRS). This is an important cause of blindness, deafness, congenital heart disease and mental retardation [1].

An estimated 238,000 children are born worldwide with CRS each year, a majority in the developing countries. The report of overall incidence of rubella immunity mother during the first three months of pregnancy is 55%, and nearly 45% of women were susceptible to CRS [2].

If primary rubella infection occurs during pregnancy, the rubella virus will cross the placenta, and induce fetal infection depending upon the time of gestation.

Infection occurring in the first 12 weeks of pregnancy causes congenital rubella infection in 90%, with almost a 100% risk of congenital defects. From 13 to 17 weeks the risk of infection is about 60%, and risk of defects about 50%. From 18 to 24 weeks the risk of infection is about 25%, with hardly any risk of congenital defects [3].

CASE REPORT

We are reporting here a case of a girl aged 4-years born to parents of non-consanguineous marriage who presented with the history of reduced hearing in both the ears since birth.

Consequently there was also delayed development of speech.

There was no history of ear discharge or ear ache.

Antenatally, the mother gave history of having developed fever with rash in the fourth month of gestation. Rash was erythematous, non-pruritic localized to the hands and feet. No history of arthralgia. This was a self limiting episode which lasted for 4 days. No history of drug intake. Rest of the antenatal period was uneventful.

This baby was delivered after Full term gestation by Lower Segment Caesarean Section. Indication for Caesarean section was cord entanglement around the neck and breech presentation. The child cried at birth.

In the post-natal period, child had difficulty in feeding and respiratory distress. During evaluation child was diagnosed to have Patent

Ductus Arteriosus (PDA).The child was treated initially for congestive heart failure. She had undergone ligation of the PDA at the age of 2 months. Presently the child is haemodynamically stable.

On general physical examination, she had microcephaly. Ocular examination revealed a coloboma in the left iris [Table/Fig-1], squint and choroidoretinal coloboma. Examination of the nose showed a broad nasal tip. Both the nasal cavities were patent and rest of the nasal cavity was normal.

Ear examination revealed bat ear on the left side. Pre-auricular, post-auricular area external auditory canal, tympanic membrane were bilaterally normal. Child did not respond to tuning fork tests and to sound clinically. All investigative parameters within normal limits except for Rubella IgM positive (1.28) (<0.80) Rubella IgG positive 162 IU (<10).

Otoacoustic emissions screening at birth was stated as PASS. Middle ear pathology was ruled out. Repeat otoacoustic emission (after 2 years): BORDERLINE.



[Table/Fig-1]: Showing left iris coloboma and broad nasal tip

BERA done at two separate instances showed NO PEAK EVEN AT 99 dB.

A diagnosis of BILATERAL PROFOUND HEARING LOSS secondary to congenital rubella syndrome was made.

With the help of Speech and Hearing department, high gain hearing aid was fitted and Audiology training and Speech rehabilitation was started. Now she is undergoing speech therapy and rehabilitation.

DISCUSSION

Rubella virus, a togaviridae, is a member of the genus Rubivirus. It is a RNA virus with an icosahedral capsid. The virus spreads by droplet infection. The primary infection is usually mild or sub-clinical characterized by a brief prodromal phase. Maculopapular rash are seen in the post-auricular area as well as over the body, accompanied by coryza and conjunctivitis, and arthralgia. It is a self limiting episode [4].

However maternal infection can transfer the infection trans-placentally and cause congenital defects in the fetus. During the period of maternal viremia the placenta may become infected causing necrosis and desquamation of the epithelium of the chorionic villi and the endothelium.

These culminate in placental hypoplasia and placentitis which in turn results in viral entry into the fetal circulation by embolic transport [5] CRS manifestations may be transient like purpura, permanent structural manifestations (deafness, central nervous system defects, congenital heart disease, cataract), or late-emerging conditions (diabetes mellitus) [6]. Hearing loss occurs in 70-90% of CRS cases, and in 50% of these children it is the only sign of CRS, which is often not detected initially. It is like that the virus gains access to the inner ear through the blood supply of the stria vascularis and cystic dilation of the stria vascularis, and partial collapse of Reissner's vestibular membrane is noted [5]. If congenital heart disease is also present, treatment with loop diuretics and aminoglycosides may add to the hearing damage.

Ocular manifestation include Pigmented retinopathy, Cataracts: pearly, dense, nuclear, Microphthalmos and Iris abnormalities [5,6], Direct viral damage of the septa of the heart may be the cause of the increased incidence of septal defects. The most common cardiovascular lesions are patent ductus arteriosus associated with infection 11 to 48 days after fertilization [5].

Demonstration of sero conversion and presence of high IgM titres is the primary mode of diagnosis of acute rubella in pregnancy. IgM in a fetus indicates intrauterine infection. IgG is the only maternal immunoglobulin that is normally transported across the placenta [7].

CONCLUSION

Congenital rubella syndrome, though uncommon nowadays, is still present in India. Presentation is often associated with subtle eye and ear anomalies. Otorhinolaryngologists should consider congenital rubella syndrome as a cause for congenital deafness more so because it can be prevented by vaccination.

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