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Rubella Syndrome associated with hearing loss and ocular defects - A Single Case Study.

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Abstract:
Congenital rubella syndrome (CRS) is an illness in infants that results from maternal infection with the rubella virus during pregnancy. When rubella infection occurs in the second month of gestation; this impairment is sensorineural. The abnormalities most commonly associated with CRS include sensorineural deafness, cardiac defects, and visual impairment.
We present a case study of 8years old, female that tested positive for the rubella virus. The research was carried out by reviewing the medical records of the case and taking the case history from the mother. The history of the mother included fever and rashes during the early gestation period (2month) which may be a possible cause of Rubella syndrome in the child. The child was born with low birth weight and as per medical reports the child was diagnosed with rubella keratopathy in both eyes, the impression of CT scan of the head was abnormal, and the impression of BERA was bilateral profound sensorineural hearing loss, however, on echocardiography, no cardiac abnormalities reported.
The characteristics of the case with CRS were observed that profound sensorineural hearing loss, intellectual impairment, ocular defects and do not have cardiac abnormalities as per medical reports. J. M. Best, 2007 also reported that clinical features of congenital rubella syndrome consist of sensorineural hearing loss, intellectual impairment, cardiac defect, and ocular defects, which support this study. Any child with bilateral or unilateral hearing loss and/or ocular defects, intellectual disability, and cardiac defects should be investigated thoroughly for CRS. Detailed case history and reviewing of medical records should be prioritized for the planning of intervention and home-based management.
Key Word: *Rubella, Teratogenic, CRS, TORCH, Hearing Loss, Ocular Defect*

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I. Introduction

Rubella virus is an RNA virus that is an enveloped single-stranded that belongs to the genus Rubivirus of the Togaviridae family. In 1985 cooper and in 2000 WHO suggested that only one type of rubella virus is described and humans are the only known host (Cooper, 1985; WHO, 2000). An ex- anthematous illness characterized by nonspecific signs and symptoms including a transient erythematous rash on the body, low-grade fever, arthralgia, and lymphadenopathy is the infection caused by this virus (Centre for Disease Control and Prevention, 1998).

Rubella infection is transmitted by droplets and is highly contagious. Rubella was endemic in many countries before the introduction of the vaccine. According to the Centre for Disease Control and Prevention in 1998 epidemics were superimposed on the endemic infection every 6 to 9 years in the United States and every 4 to 5 years in Europe (Centre for Disease Control and Prevention, 1998). Teratogenic effects of primary rubella infection in pregnant women have the major public health importance of rubella infection (Gregg, 1941; South and Sever, 1985). It appears to be of crucial importance at which stage of pregnancy disease is developed. If the infection in the early months of pregnancy may result in multiple congenital fetal abnormalities that are known as congenital rubella syndrome (CRS).

The CRS shows various abnormalities including sensorineural deafness, cardiac defects (ventricular septal defects, patent ductus arteriosus, peripheral pulmonary artery stenosis, and hypoplasia or coarctation of the aorta), visual impairment (ex; cataracts, glaucoma, pigmentary retinopathy, chorioretinitis, and microphthalmia),

and CNS pathology (ex; intracranial calcifications, microcephaly, behavior disorders, and meningoencephalitis mental retardation). In addition, infants exhibit both intrauterine and postnatal growth retardation infected with CRS. According to the Centers for Disease Control and Prevention in 1998, Women who acquire rubella during the first 20 weeks of pregnancy have been estimated to occur among 20-25% of infants (Centers for Disease Control and Prevention, 1998).

J. M. Best did a study on 'Rubella' in which he found that gestational age at the time of confirmed rubella in pregnancy and clinical manifestations of congenital rubella syndrome, which can be found below.

Clinical manifestation	**gestational age (weeks)**
Deafness	2–18
Cataract	3–12
Congenital rubella retinopathy	2–18
Cardiac defects	3–13
Neurological defects	3–16
Multiple defects	3–12
No defect	7–>20

From the above table, it is observed that Clinical manifestations of congenital rubella syndrome occur at specific points during gestation, i.e., 2-18 weeks.

Rubella babies associated with the commonest abnormalities are hearing impairment, either alone or in combination with other defects (Cooper, 1975). There is a high risk of sensorineural hearing loss when infection occurs before the 18th week of pregnancy [Munro ND 1987]. Deafness is the most common abnormality seen in CRS, and the proportion of children may be as high as 70%who have hearing loss. Some of the studies, in which suggested that the contribution of congenital rubella may be higher than suspected based on clinical studies only which were done on children with congenital hearing loss. In Finland, a serological survey suggested the presence of rubella-specific antibodies in 45% of those studied in which 128 infants aged 6 months to 5 years were taken with moderate or severe congenital hearing loss (Ojala et al, 1973). It was also reported that in Trinidad 74% of children with congenital deafness had rubella antibodies (Karmody, 1968). If maternal infection occurs up to the nineteenth week of pregnancy then the child may be born with congenital hearing impairment (Cutts et al, 1997). Several studies indicate that when rubella infection occurs in the second month of gestation then the chances of having a hearing impairment are severest; this impairment is sensorineural (Goodhill, 1950; Barr and Lundstrom, 1961; Anderson et al, 1970).

Ocular defects, a "salt, and pepper" retinopathy is seen in 40– 60% of cases and also most commonly observed but does not typically have visual consequences unless neovascularization develops (M. L. Tamayo et al, 2013). The retinopathy is most commonly located in the posterior pole and is also varied in appearance and distribution. Nuclear cataracts are bilateral in 50% and occur in about one-third of all cases (R. S. Duszak, 2009, J. M. Best, 2007). Other diverse but less common abnormalities have been reported (M. A. Menser and R. D. K. Reye, 1974). Maternal rubella infection at different stages of pregnancy will lead to different rates of fetal infection and also combinations of classic CRS features.

II. Case Discussion:

Case Name-A Age/Sex-8 Years/Female

P. Diagnosis-Congenital Rubella Syndrome with Hearing Impairment

Complaint-Child has hearing difficulty in both ears, inability to speak age-appropriate and weak eyesight as reported by her mother.

Medical History:

Pre-natal: mother had a high fever and rashes all over her body during two months of pregnancy. Mother was unaware that she was two months pregnant so; she took medicine for rashes and fever as a consultant by the doctor. But mother was not taking treatment properly by expert doctors due to a lack of knowledge. In study by M.M. Rahman et. al., showed that the presence of antibodies in the children correlated well with the presence of antibodies in the mother and also with the maternal history of rash and fever during early pregnancy

Perinatal: Delivery was full term (9month) and normal at the hospital. The weight of the child at the time of birth was less than 2Kg as reported by her mother. The birth cry could not be reported by her mother.

Post-natal: The child was suffering from fever after 8days of birth as reported by her mother.

The mother reported that the child had vision impairment in both eyes, so ultrasound of the eyes was recommended by the Doctor at KGMU. Ultrasound of both eyes was done at KGMU, Lucknow, UP on 03rdApril 2013.

Impression- Bilateral normal posterior segment

After that, her mother consulted with doctors of Shroff Charity Eye Hospital (SCEH). They recommended the [toxoplasmosis, rubella, cytomegalovirus, and herpes] TORCH Test on 30th July 2013.
TORCH Profile:
Anti- Rubella IgG is more than 350 IU\ml (positive)
Anti-Rubella IgM is more than 1.08 IU\ ml (positive)
Echocardiography was done on 11th Feb 2014.
Impression- Normal Cardiac Chamber
LVEF is 60%
No valvular lesion is seen
No PE/ thrombus/Mass is seen
CT Scan of Head P\C done on [11\02\2014]
Impression- Gyri from calcification in bilateral frontoparietal regions with abnormal leptomeningeal enhancement.
Surgery of both eyes was done at Dr. Shroff Charity Eye Hospital on 12 April 2014
Diagnosis: Rubella keratography
As the child had not shown any such improvement in communication, the mother of the child was concerned about her speech and hearing skills and consulted doctors at T S Mishra Medical College (TSMC), Lucknow and further hearing testing was done at TSMC On 05 February 2016.
Behavioural Response: Auro-palpebral reflex (APR): Absent
Arousal test (Awakening): Absent
Auditory Brain Stem Response Findings: Testing at intensity level of 100dB nHL, both ears indicated absolute and interwave latencies were mild prolonged. Well identifiable V peak were unclear at decreasing down to 90 dB nHL. Unclear morphology seen below 90 dB HL
Interpretation-B/L Profound SNHL
After hearing testing, the child used bilateral BTE body level hearing aid till the age of 7 Yrs. but the mother did not notice any improvement in the auditory behavior of the child.
Therefore, the parents again consulted their doctor/s of TSMC and the doctor recommended cochlear implant surgery at Sanjay Gandhi Postgraduate Institute of Medical Science (SGPGI), Lucknow.

Impressions of Hearing testing done at SGPGI:
OAE- Refer in both the ears.
BERA- No identifiable waveform was seen on the right side both at 95 and 75 dB. No identifiable waveform was seen on the left side both at 75dB and 95dB.
FFA- Profound SHNL in both the ears.
Cochlear implant surgery in the right ear was done on 17 January 2021 at SGPGI, Lucknow. The further child uses body level hearing aid in the left ear. After healing of surgery doctors of SGPGI recommended and referred the child for IQ Test at King George Medical College (KGMC) Lucknow.

IQ Test was done on 12 July 2021at KGMC, Lucknow:
Results: Social age-7 Years 10 Months and Social quotient- 90-94
The result shows speech production deficits and comprehension is intact.

Language status
The child is using some speech sound \a\ \p\ \m\ \b\ and after cochlear implant surgery and child is saying \amma:\ to her mother, \ pa: pa:\ to her father occasionally as reported by his mother.
REELS were administered: RLA: 30 – 33 Months and ELA: 11 – 12 Months

Listening status
Child listening status is not fair. The child is responding to loud sounds always like crackers, vehicle horns, and sometimes shows a response on calling her name as reported by her mother, and the response is noticed by eye blinking, cessation of activity, head turn, and shocking. The child shows no response at a comfortable level and low-intensity level sounds.
Awareness/Detection: the child is aware of the sound of crackers, vehicles horn, drum beat, and sometimes shows response on calling her name.
Discrimination: The child can discriminate her mother and father sounds.
Identification: The child can identify her mother, father, and brother sounds.
Comprehension: The child is not able to respond to several kinds of verbal request

III. Plan

Auditory training;
1) Awareness: focus on developing listening skills. The child will be responding to different low frequency (drum,\a\\m\ \u\ \i\ and high frequency (whistle, horn, plate, bowl,\s\ \sh\) auditory stimuli at a distance of 2feet, 5 feet and 8 feet.
2) Discrimination: child will be able to discriminate sound of low frequency (drum, \a\\e\\u\\m\), high frequency (horn, whistle, plate \s\ \sh\) sounds. The child will also be able to discriminate animal sounds like dog and cat using minimal prompts.
3) Identification: the child will able to identify the sounds of animals like dogs and cats and different auditory stimuli of low frequency (drum \a\ \e\ \u\ \m\)and high frequency (whistle, plate, horn,\s\ \sh\) by using minimal prompts.
4) Comprehension: the child will be able to comprehend phrases level using visual cues (ex; subject + verb+ object)
5) Speech: focus on production of \a\ \i\ \u\ \m\ \s\ \sh\\p\ \b\ speech sounds.
6) Language:
Receptive language: Identification of her common things (eg.bag, pen, pencil, bottle), fruits (ex; apple, mango, banana) and animals (ex; dog, cat, cow).
Expressive language: Production of \pa: pa:\ \mamma:\ \bha:i\, fruits name (apple, mango, banana), animals name (dog and cat) and common things (bag, pen, and copy)
Goals were also planned by the other professionals (i.e., Physiotherapist, Occupational therapist, Special Educators) to improve her vision ability, motor skills, and cognitive ability at CRC, Lucknow.

IV. Discussion:

In a study by SA Plotkin et al. 2011, maternal rubella infection usually occurs 5 to 7 days after maternal inoculation. The virus spread across the placenta hematogenously, leading to congenital infection in developing fetus including miscarriage, stillbirth, and various teratogenic effects. In another related study done by M.M. Rahman et. al (2003). It was found that the presence of antibodies in the children correlated well with the presence of antibodies in the mother and also showed a maternal history of rashes and fever during early pregnancy. In our study rubella infection may be spread during 8 weeks (2 months) of gestation period in a developing fetus which caused teratogenic effects. Also, the mother had a fever and rashes all over the body but she was unaware of rubella infection due to lack of knowledge, so proper treatment was not taken to her.

In our study case was diagnosed with rubella keratopathy in both eyes (it is a type of ocular defect) which might be a sequel of either associated glaucoma or direct viral cytopathologic effect. However, a study was done by. Vijayalakshmi (2002) in which he took 46 seropositive infants of 12 months of age. The result found that 41 patients had both eyes affected.

J M Best did a study in 2007 in which he found out that clinical features of congenital rubella syndrome consist of sensorineural hearing loss, intellectual impairment, cardiac defect, and ocular defects. However, in our case with congenital rubella syndrome, characteristics found are profound sensorineural hearing loss, intellectual impairment, ocular defects (rubella keratopathy in both eyes) and do not have any cardiac abnormalities according to medical reports.

In a study of LPA Silva da et al 2006, congenital rubella was the cause of hearing loss in 32% of patients with deafness and OA Lasisi et al, did a study in 2006, in which it was found that rubella was considered to be one of the causes of sensorineural hearing loss. In our study, the case has bilateral sensorineural hearing loss which is probably caused by rubella syndrome.

V. Conclusion:

The mother of the child was concerned about her hearing and speech problems of the child so she went to T.S Mishra medical college (TSMC) for consultation. The child was diagnosed with profound SNHL. She got recommended and also got fitted with a body level hearing aid by the audiologist at TSMC. After fitting of hearing aid mother did not notice any improvement in the auditory and communication skills of the child. So, she was referred from TSMC to SGPGI Lucknow for a cochlear implant (CI). CI surgery was done at SGPGI Lucknow on 17 January 2021. After 1 year of CI, the mother again did not notice any kind of improvement in auditory and communication skills. Then she came to CRC Lucknow on 18 December 2022 for further assessment and intervention. The child was assessed and medical records of the child were reviewed by the SLP and other rehabilitation professionals of CRC. It was found that the child is having a problem with profound SN HL, Vision problems, Intellectual disability, and issues with motor skills. Accordingly, the intervention was planned for a better prognosis for the child. At present, Mother is satisfied with the intervention plan, which also indicates that a detailed case history and reviewing of medical records by the team of rehabilitation professionals should be prioritized for the planning of intervention and home-based management.

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Conflict of interest: Nil

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Written consent was taken from the mother to conduct the case study and present the findings.

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