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Vishwanath Hesarur*

Assistant Professor, Department of Cardiology, KLE University's Jawaharlal Nehru Medical College, KLES, Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi, Karnataka, India

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***Corresponding author:** Vishwanath Hesarur, Assistant Professor, Department of Cardiology, KLE University's Jawaharlal Nehru Medical College, KLES Dr Prabhakar Kore Hospital and Medical Research Centre, Belagavi, Karnataka, India, Tel. No: +919480008361; Email: drvishwanathesarur@yahoo.com

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Case Report

A Rare Case of Congenital Rubella Syndrome

Abstract

Introduction : Congenital rubella syndrome (CRS) is a consequence of rubella infection that can occur when the virus is transmitted in utero during maternal primary infection. It still affects 110,000 children around the world. It has a wide spectrum of presentation which ranges from silent viremia to spontaneous abortions, blindness, deafness, congenital heart disease, and mental retardation.

Case Report: Here, we report a case of CRS in a girl aged 8 years who presented with congenital cataract, severe post ductal coarctation of aorta with patent ductus arteriosus and bilateral sensory neural hearing impairment.

Conclusion: We present a case report of congenital rubella syndrome (CRS); though uncommon nowadays, it is still present in India. The main characteristics of our case report were congenital cataract, severe post ductal coarctation of aorta with patent ductus arteriosus and sensory neural hearing impairment. This case is reported to highlight the importance of vaccination by which rubella infection can be eradicated.

Introduction

Congenital rubella syndrome (CRS) is a consequence of rubella infection that can occur when the virus is transmitted in utero during maternal primary infection. It still affects 110,000 children around the world [1]. It has a wide spectrum of presentation which ranges from silent viremia to spontaneous abortions, blindness, deafness, congenital heart disease, and mental retardation [2]. Here, we report a case of CRS in a girl aged 8 years who presented with congenital cataract, severe post ductal coarctation of aorta with patent ductus arteriosus and bilateral sensory neural hearing impairment. This case is reported to highlight the importance of vaccination by which rubella infection can be eradicated.

Case Report

A 8 year old girl presented to our OPD with history of exertional breathlessness and palpitation of 2 months duration with a background history of congenital blindness of right eye and decreased hearing since birth. Her past history revealed that she was born out of a non-consanguineous marriage, full term normal vaginal delivery at Government hospital. Baby cried immediately at birth. Weight at birth was 2.5kg. There was no history of chronic illness in her parents, though the mother had an episode of fever in the first trimester of pregnancy, there was no history of rash during that episode.

There were no developmental delays in achieving motor and social milestones; however there was delay in achieving speech output. Her head circumference was normal at birth and the treating doctor could not diagnose CRS at birth. No virologic studies were done previously. Since both parents are uneducated and from village, were not aware of vaccination against rubella.

On general physical examination

She was undernourished, with a low weight for age, had micrognathia, right eye nuclear cataract with a marked eye asymmetry, horizontal nystagmus of the right eye and a broad nasal tip (Figure 1).

She was acyanotic with a saturation of 96% at room air. She had a radio-femoral delay but no radio-radial delay. There was a grade 3/6 harsh pansystolic murmur best heard in the left infraclavicular area. Rest of the examination was unremarkable. She did not respond to sound clinically and was subjected to brainstem evoked response audiometry (BERA).

Her echocardiographic examination revealed situs solitus, severe post ductal coarctation of aorta with a peak pressure gradient of 50 mmHg, a small patent ductus arteriosus with only systolic flow from the aorta to the right descending pulmonary artery and a bicuspid aortic valve (Figure 2).

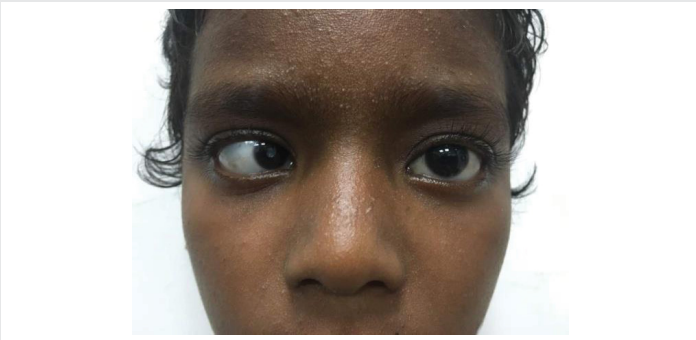


Figure 1: Photograph showing micrognathia, right eye nuclear cataract with a marked eye asymmetry, horizontal nystagmus of the right eye and a broad nasal tip.

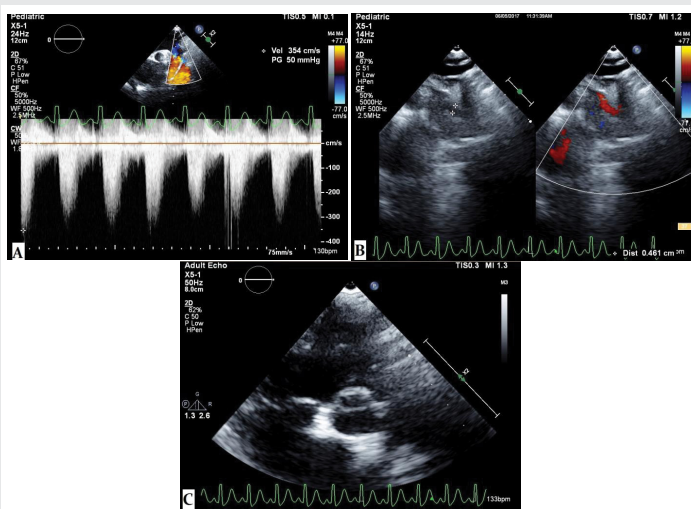


Figure 2: Transthoracic echocardiography showing severe post ductal coarctation of aorta with a peak pressure gradient of 50 mmHg (2A), a small patent ductus arteriosus (2B) and a bicuspid aortic valve (2C).

Investigations

Her hematological parameters, renal, Coagulation and thyroid function tests were within normal limits. Her Rubella IgM positive (1.28) (<0.80), Rubella IgG positive 182 IU (<10).

Discussion

Rubella is a togavirus belonging to genus rubivirus. Rubella is usually a mild febrile rash disease in children, but to young women's pregnancy, especially in their first 16 weeks, it has devastating consequences [3]. Both deafness and cardiac manifestations occur if viremia takes place before 11 weeks, and viremia between 13-16 weeks results in isolated deafness [4]. Infection occurring in the first 12 weeks of pregnancy causes CRS in 90%, with almost a 100% risk of congenital defects. From 13 to 17 weeks, the risk of infection is about 60%, and risk defects about 50%. From 18 to 24 weeks, the risk of infection is about 25%, with hardly any risk of congenital defects [5]. Though the exact teratogenic mechanism of in utero rubella infection remains unidentified, viral replication in the cells during fetal organogenesis seems to be accountable for CRS [6].

The first description of CRS belongs to Gregg in 1941 but it was completely described in 1944 [7]. A study of Miller et al. showed that the risk of congenital infection was 81% and the risk of malformation was of 69% if the mother had rubella in the first pregnancy trimester. The risk fall to 33% after 12 weeks of gestation and no defects were encountered after week 16 [8]. Intrauterine growth retardation and prematurity frequently manifest in CRS. The commonest defect is central hearing loss. Characteristic cardiac defects that occur in CRS include PDA, pulmonary stenosis, and ventricular septal defect (VSD). Cardiac lesions are more frequently prevalent in neonates with ocular lesion [2,9].

There are still congenital rubella syndrome cases reported around the world despite the introduction of rubella vaccine in the national immunization programme of many countries [3].

Conclusion

We present a case report of congenital rubella syndrome (CRS); though uncommon nowadays, it is still present in India. The main characteristics of our case report were congenital cataract, severe post ductal coarctation of aorta with patent ductus arteriosus and sensory neural hearing impairment. This case is reported to highlight the importance of vaccination by which rubella infection can be eradicated.

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