

CASE REPORT

A Neglected Congenital Rubella Syndrome in 10-Year-Old Boy

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Abstract

Introduction: Cataract, congenital heart defect, and hearing impairment are the three most common manifestations of congenital rubella syndrome (CRS) that lead to developmental delay in the children. CRS usually present during the infant period of three months old as the median age number. Ironically, we found a 10-year-old boy who was diagnosed with CRS and there was no significant plan to improve his condition anymore because of his neglected by family. The purpose of this case report is to alert primary health workers and also ophthalmologists to do our best to prevent blindness due to cataracts in Indonesia with early detection. **Case presentation:** A 10-year-old boy was brought to the outpatient clinic with a chief complaint of difficulties in focusing the target in the classroom. The grandmother said that her grandson has already had a white plaque in his both eyes since he was born. The boy has also suffered from deaf-mutism. His visual acuity was hand movement and there was lens opacity in his both eyes. He also had posterior uveitis in both eyes followed by the presence of nystagmus. There was retinal detachment in ultrasonography (USG) examination. The patient has no cardiac abnormalities. Rubella infection was confirmed by the high number of IgG rubella serology. His grandmother had just brought the boy to the outpatient clinic because she never thought that the eyes needed therapy until the boy could not do his daily activities as the cataract worsen. **Conclusions:** The parent's role is highly crucial for the visual outcome of patient with congenital ophthalmologic problems. Primary health worker also play a significant role in early detection that may prevent such incident. Delayed detection and management of congenital cataracts may cause permanent visual impairment and lead to patient's poor quality of life.

Keywords: neglected; rubella; cataract

Introduction

Rubella infection during the first trimester of pregnancy may cause several fetal malformations known as congenital rubella syndrome (CRS). Several birth defects may present from rubella infection, despite today's massive worldwide vaccination efforts. During 1963–1965 there was rubella epidemic that affected 10% of all pregnant women worldwide.^[1] According to World Health Organization (WHO), 70% of world's infants had already received rubella containing vaccine (RCV) in 2020, yet there were still a few of those who did not and resulting in some serious conditions.^[2]

Ocular, otologic, and cardiac abnormalities followed by microcephaly and various developmental delay are the most common consequences of Rubella infection.^[3] Ocular consequences of CRS are not limited to abnormalities noted in the neonatal period. Additional abnormalities may appear years and even decades after birth.^[4] The neglected in congenital ocular disorder may lead to several serious consequences including learning impairment and also developmental delay. Delayed treatment in CRS resulting in some serious complications that affect the patient's quality of life.^[5] This patient could not communicate with anyone at all because of his blindness and deaf-mutism. If it was managed earlier, these children might have the possibility of having best visual acuity.^[6] The

purpose of this case report is to make all primary health workers aware of the importance of early detection in congenital ophthalmology problems. If it was found and referred in a timely manner to more advanced professional healthcare needed, they might have a better prognosis and definitely a better quality of life.^[7]

Case presentation

A 10-year-old boy was brought to the outpatient clinic with a chief complaint of difficulties in focusing the target in a classroom. The teacher said that he always did his activities at a very close distance in special education school. According to his grandmother, there was a white appearance on both eyes since birth. The whitish left eye was denser than the right eye. They never went to a doctor since the patient was still able to do his daily activity. Shortly after born, his parents left him so and the boy was raised by his grandmother for the past ten years. He had a history of deaf-mutism and developmental delay. The patient used sign language as a daily conversational method. There was no redness, secrete or pain in both eyes.

There was no abnormality during pregnancy and they never went to a doctor. The patient's birth weight was 3.000 g and 48 cm in length with normal delivery history. The vaccination record was uncertain since his grandmother was not very well educated as if for the breastfeeding and nutritional history. There was no history of trauma, nor any other surgical procedure done by the patient.

The grandmother said that as the patient grew up, he could communicate using sign language since the cataract in the right eye was not as dense as in the left eye. However, now, the right eye's cataract was quite dense with the appearance of peculiar nuclear cataract (Figure 1). The patient could not do any activity nor communicate with anyone even his grandmother because of his deaf-mutism as well.

Physical examination showed that the visual acuity was hand movement in both eyes and there was a presence of nystagmus at his first came. Retinal detachment was found in the right eye during the ultrasonography (USG) examination. The posterior segment could not be evaluated because of the dense bilateral cataract

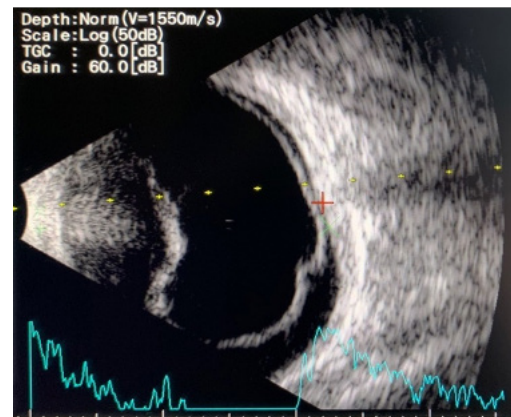


Figure 2. USG Examination in the right eye showed a membrane echogenic lesion with 100% RCS complex, giving an impression of a retinal detachment.

(Figure 2). Laboratory findings showed that there was a past rubella infection which was shown the IgG Rubella serology test. The pediatric audiology department concluded that there was no organic abnormality in both ears so this patient was diagnosed with congenital sensorineural deaf-mutism. Echocardiography done by pediatric cardiologist showed the normal function of his cardiac condition.

This patient was diagnosed as a confirmed case of congenital rubella Syndrome by the fact that he had two out of three most common findings of CRS patients (cataract and sensorineural deaf-mutism) even though he had no cardiac defect, with addition of two-fold increase in the number of IgG serology test.^[8] Examination under general anaesthesia and simple lens aspiration followed by IOL implantation of his right eye was done in order to have a better visualization through the posterior segment. Unfortunately, there was no improvement in post-operative visual acuity in both eyes. At the post-operative care in the outpatient clinic, a definite vitreous haze and retinal detachment with demarcation line in the temporal area were seen in his right eye (Figure 3).

Discussion and conclusions

Congenital cataracts play a significant role in causing worldwide children's vision loss with nearly 10% of all blindness cases. It is estimated that there are 1 to 6 congenital cataracts cases per 10.000 live births.^[9] Delay in diagnosing children with congenital cataracts may

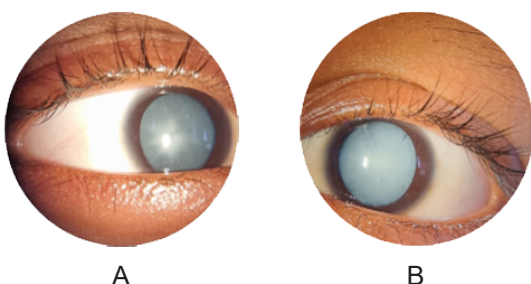


Figure 1. Slit lamp examination: Diffuse light; (A) Right eye, (B) Left eye. Both photos showed a peculiar nuclear cataract.

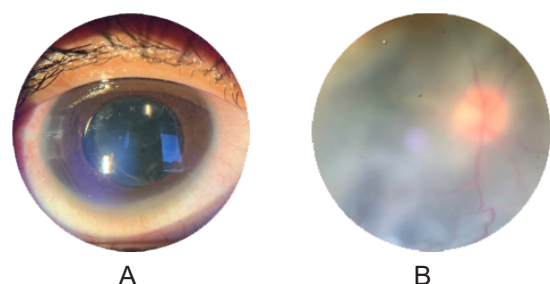


Figure 3. (A) Right eye's post-operative photo; (b) Right eye's fundus photograph showed vitreous haze and retinal detachment.

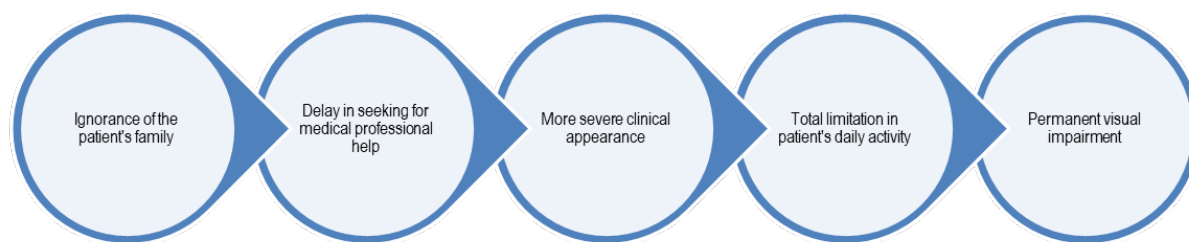


Figure 4. Schematic process of this patient's delayed treatment.

result in some serious complications which affect the patient's quality of life, although several aspects might cause the delay (Figure 4).^[7]

There was no doctor visit throughout these past ten years because his grandmother thought that the limitation in the patient's activities was normal in children with deaf-mutism. It showed that his grandmother was somehow neglected the fact that her grandson was not fine. The patient did all of his daily activity only by using his right eye since the cataract in his right eye was not as dense as in the left eye. Since the right eye became denser, the patient did all of his daily activities with the help of his grandmother.

The presence of nystagmus in this patient proves that cataracts might be bilateral from the beginning. Although the right eye's cataract was not as dense as the left one, the possibility of developing visual deprivation amblyopia is very likely.^[10] Bilateral lens opacities generally tend to cause less severe vision deficits than unilateral cases, because interocular competition in unilateral cases adds to the direct impact of image degradation. The right eye must be the priority in performing cataracts surgery, although ideally, for optimal visual development, a bilateral dense congenital cataract should be removed within the first ten weeks of life.^[3]

The grandmother's leading cause of the delay in seeking medical professional help was ignorance regarding birth defects, especially in a developing country as Indonesia (Figure 4). This process resulted in a more severe clinical appearance shown by the limitation of the patient's daily activity since the patient also has congenital deaf-mutism, whereas before, the patient could still do his daily activity such as eating and taking a shower by himself. All of these neglected processes with the addition of retinal detachment, result in a permanent visual impairment.^[11]

CRS can be diagnosed with a triad of ocular manifestations, auditory, and cardiac defects.^[10] The approved definition of a confirmed CRS case by the Council of State and Territorial Epidemiologists (CSTE) is the presence of at least one of the symptoms that is clinically consistent with rubella such as cataracts, congenital heart disease, hearing impairment, hepatosplenomegaly, developmental delay, and microcephaly with the addition of laboratory evidence of rubella infection.^[8]

This patient fulfilled two out of three most common signs of congenital rubella infection which were cataracts and sensorineural hearing impairment that lead to his mutism and developmental delay with the addition of laboratory findings showed a two-fold increasing number of IgG rubella tests. The quality of life in patient with congenital ophthalmologic disease is based on several factors such as the density of cataracts, bilateral cataracts, surgery process and visual acuity. The most important factor that affects a patient's quality of life is visual acuity.^[12] Since this patient had already lost his ability to see, hear, and speak without any cardiac abnormalities or any other systemic problems, we can conclude that his quality of life might be very poor.

The neglected by family of this patient's condition plays a major role in terms of time whether the poorly educated parents seek medical professional help. Ironically, the neglect resulted in a poor prognosis visual outcome which affected the patient's quality of life. In conclusion, primary health worker plays a significant role in early detection that we might prevent such incident. Delayed detection and management of congenital cataracts may cause permanent visual impairment. Blindness and sensorineural hearing impairment that leads to his inability to speak conclude that his quality of life would be poor. This might be just a tip of an iceberg of the big picture of Indonesia's cataracts problems. As an ophthalmologist, we should do our best not only to educate the patients about congenital ophthalmologic problems but also to form a thorough system for its early detection to be able to prevent this incident.

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