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Managing a Child with Acute Rheumatic Fever in a Remote and Rural Clinic: Role of Decentralized Primary Healthcare

Acute rheumatic fever (ARF) is an autoimmune inflammatory process that develops as sequelae of Group A streptococcal (GAS) infection. Most cases of ARF occur in children aged 5–15 years. Apart from the fever, ARF has extremely variable manifestations, with heart and joints being commonly involved. In a small proportion of children, especially in girls, the basal ganglia may be affected. This manifests as chorea or hemichorea. Although no specific diagnostic or confirmatory test exists, a combination of symptoms, signs, and supportive tests (Modified Jones criteria) is applied to help establish diagnosis. Persons affected with ARF are predisposed to recurrences following subsequent GAS infection, resulting in the dreaded cardiovascular sequelae rheumatic heart disease (RHD).

In this article, we share a case report of a 13-year-old girl who presented to one of our rural primary health-care clinics with abnormal, purposeless movements of one-half of the body. Based on her presenting signs and symptoms, we diagnosed her as having ARF, and managed her accordingly. Using this case report, as an illustration, we discuss the challenges of diagnosing and managing ARF in rural primary health-care settings, and various solutions that theoretically exist for the prevention of RHD but need to be practiced in ground reality on a much wider scale.

CASE REPORT

Sarita (name changed), a 13-year-old girl presented to the AMRIT Clinic at Rawach, a remote village, among the last villages of the hilly region of Mewar. She was brought to the clinic by her parents. Sarita's complaints were primarily the insidious onset of abnormal movements of her left upper limb for around 1.5 months. Due to the movements, she was having difficulty in walking and carrying out activities of daily living. She had stopped going to school since the movements had started. On probing further, we elicited a significant history of having painful swelling of knee joints initially, followed by similar involvement of ankle joints a few days later. The swelling was not associated with restriction of movements. We also learnt that Sarita has been having mild grade fever since the onset of illness. Her parents had consulted various traditional practitioners and received some medications. Her fever and joint swellings had resolved a week or two before the limb movements had started. There was no history of preceding sore throat or skin lesions.

She had not experienced similar complaints in the past, and neither had any other family member. Her parents are farmers who own a small piece of land that they maintain for their livelihood. Neither of the parents ever went to school. Sarita

is the third child in birth order. She has two sisters and an elder brother; who works as a migrant worker, providing financial support to the family. They live in a single room *kutchra* house with poor ventilation.

On arrival, the child was conscious and oriented, but irritable. She was having continuous, purposeless movements of the left upper limb and to a lesser extent of the left lower limb, indicative of chorea [Video 1]. There were no other neurological abnormalities or deficits.

Sarita had a temperature of 99.9°C with elevated pulse rate of 100/min, that was regular, with normal volume and character. The respiratory rate was 20/min, and the blood pressure was normal. The jugular venous pressure was not elevated. The child had mild pallor and no cyanosis. There was no edema or other signs of congestive heart failure. The skin did not show any nodules or rash. Her joints were normal, without any swelling, erythema, or tenderness, and displayed normal range of movements. The salient examination findings on systemic examination were the absence of cardiomegaly and a grade 2 soft diastolic murmur in the mitral area on cardiac auscultation. The liver and spleen were not palpable, and the remaining systemic examination was normal.

Based on the presence of three major Jones Criteria – chorea, carditis and migratory polyarthritis- we made a probable diagnosis of ARF.^[1] However, we faced major challenges, when it came to satisfying the essential criteria. We could not test for antistreptolysin O (ASO) and erythrocyte sedimentation rate, because the child was extremely uncooperative (*see later*), and would not allow us to draw blood for a sample. For the same reason, we could not take an electrocardiogram. In addition, the parents were unable to take the child to a city hospital for an echocardiography (ECHO), since it is situated about 100 km away.

Sarita displayed extreme fear of all medical procedures. When we tried to give her injectable Benzathine Penicillin (BP), she ran away from the clinic. And, when staff members and her parents attempted to bring her back, she began to throw stones at them to ward them off. However, with some patience and counseling, she agreed to return on the condition that she is given oral medication, instead. She was started on oral amoxicillin (50 mg/kg for 10 days) for controlling the active infection, and sodium valproate (20 mg/kg) for managing the chorea. We also added prednisolone (1 mg/kg) for managing the presumed activity.

On follow-up, after a week, her irritability had reduced and chorea lessened significantly. Vitals were stable, but tachycardia was still persisting. A well-audible diastolic murmur in the



Figure 1: Sarita with Senior Health Worker of AMRIT Clinic 3 weeks after initiating treatment

mitral area had become apparent, with no opening snap. The symptomatic improvement and establishment of rapport with staff members resulted in our managing to convince Sarita to submit to at least one shot of intramuscular 12 lacks units of BP after a drug sensitivity test. Once she had experienced it, she overcame her morbid fear and agreed to continue with injectable BP prophylaxis, in addition to the steroids and valproate. By the 3rd week of initiation of therapy, the chorea had disappeared, her pulse rate had decreased to 84/min and the murmur had disappeared [Figure 1].

The family was satisfied with her recovery. A month afterward, Sarita did not come for her scheduled follow-up visit. This prompted our health workers to make a home visit. It was found that the child had resumed going to school. The family was counseled regarding the need for regular prophylaxis and the consequences of missing even a single dose. She was brought to the clinic on the next day and she was administered the missed dose. Tapering off steroids was started as per standard protocol. Additional counseling was done regarding maintaining dental hygiene. The child has been under our follow-up for 3 months and is doing well.

DISCUSSION

The occurrence of ARF in developed countries has declined dramatically over the past decades, largely on account of improvement in living conditions and socioeconomic status; and early diagnosis and management of streptococcal sore throat infections.

In India, we do not have data on the incidence of ARF, but the prevalence of its sequelae, RHD, is available. Forty percent of the global burden of RHD is estimated to be from India, alone. In a review of school-based surveys conducted between 2008 and 2014, based on echocardiography results, RHD prevalence was 5 to 51 per 1000 children in the age group of 5–15 years, across different states.^[2] As expected, the states with more developed primary health-care systems such as

Kerala, had a much lower prevalence (5.84/1000 children in Trivandrum) than those with less developed health systems such as Rajasthan (51/1000 children in Bikaner district).^[2]

RHD significantly affects the quality of life, and managing RHD is a complex and expensive process; often becoming unavailable and inaccessible for poorer populations. For example, the price of a valve replacement in India ranges from Rs 300,000 to Rs 500,000. While in the long run, improvement in living conditions (and reduction in overcrowding) would lead to a decline in the incidence of ARF and RHD, there are two specific interventions that can reduce the morbidity and the burden associated with the initial illness and its aftermath.

First, timely identification and adequate treatment of an upper respiratory infection (URI) caused by Group-A streptococcus is critical. A Cochrane review of the accuracy of rapid antigen detection tests (RADts) in the diagnosis of streptococcal pharyngitis revealed fairly high sensitivity (86%) and specificity (95%).^[3] However, the cost and challenges in availability of the RADts in most primary health-care settings, restricts its use. Thus, primary health-care providers have to rely on clinical guidelines such as the Integrated Management of Neonatal and Childhood Illnesses, according to which antibiotics are not recommended for the treatment of URI. Alternately, health-care providers may use antibiotics ad hoc for inadequate duration, which do not eliminate the streptococci. In both scenarios, the risk of ARF and subsequent RHD is not diminished, underscoring the need for such a test.

Second, it is equally important to identify and manage ARF on time (as we did in this case, despite the numerous challenges), so as to prevent the progression of carditis. With suitable training, use of standardized protocols, and availability of teleconsultation (in places where medical professionals are unavailable), it is not difficult to identify and manage ARF even in a remote and rural setting, as this case study demonstrates. Non-availability of ECHO in the primary care settings, or even at district levels, makes it more difficult to timely identify, and manage ARF and RHD. Wider availability of tele-echocardiography can offset this constraint at the primary care level.^[4]

Finally, the prevention of recurrence requires that the affected individual returns every 3–4 weeks to the health facility for BP prophylaxis for a minimum of 10 years.^[5] Children belonging to marginalized populations or living in the periphery can avail these injections regularly, only if the health-care facility is close by and well stocked and equipped. We need to ensure that this essential drug is available at all Primary Health Centers and Health and Wellness Centers with requisite protocols for administration in place.

We hope that Sarita will be able to continue with the required prophylaxis. In her circumstances, there are several social barriers that may preclude this eventuality: limited mobility of a girl, dependence on male members to bring her for her

follow-up visit, and though she is only 13-year-old, the very real possibility of early marriage.

People in rural areas face many barriers to accessing quality healthcare for preventing, promoting, and treating illnesses such as ARF. Only a responsive primary health-care service with empathetic and skilled providers, can offset these challenges, and ensure requisite care. When supported with simple and appropriate diagnostics, protocols, and teleconsultation, the healthcare of populations living in rural areas, can be transformed to be available at a cost that these communities will be able to afford. It is imperative for pediatricians to support the development of such decentralized health-care systems.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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