

Sydenham's chorea in a female child with rheumatic fever in Bhutan: a case report

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Title Page

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ABSTRACT

Sydenham's chorea is a major manifestation of Rheumatic fever. It occurs primarily in children and is seen rarely after the age of 20 years.

We describe a 16 year old girl who presented with purposeless involuntary movements of upper and lower limbs. Laboratory blood reports showed raised Erythrocyte Sedimentation Rate and Anti-streptolysin O. 2D Doppler Echocardiography confirmed sub-clinical carditis, thickened mitral and aortic valve with mild mitral regurgitation. She was managed as Acute Rheumatic Fever with oral Phenoxymethyl penicillin and carbamazepine.

Early diagnosis is a key to prevent the late consequences of Acute Rheumatic Fever and Rheumatic Heart Disease.

Sydenham's chorea is a rare presentation of Acute Rheumatic Fever. Absence of clinical carditis does not rule out carditis. Echocardiography should be done.

Keywords

Sydenham's chorea, Acute Rheumatic Fever, Rheumatic Heart Disease, Sub-clinical carditis, Diagnostic criteria

INTRODUCTION

Sydenham chorea represents the most common cause of acquired chorea in childhood [1]. It is one of the major criteria for the diagnosis of acute rheumatic fever [2]. In Sydenham chorea there is both neurological

abnormalities and psychiatric disorder. The neurological abnormalities comprise of involuntary choreatic movements, incoordination of voluntary movement, muscular weakness and hypotonia [3]. Psychiatric disorders include emotional lability, hyperactivity, distractibility, obsessions and compulsions [2, 3]. Choreatic movements are involuntary, irregular, purposeless, non-rhythmic, abrupt, rapid and unsustained. It disappears in sleep [2].

The most common age for the onset of acute rheumatic fever is 5 to 15 years old [1]. Rheumatic fever is the major cause of acquired heart disease in children and up to 60% of people who present with Sydenham chorea develops rheumatic heart disease [2]. Rheumatic fever is an acute non-suppurative inflammatory complication of group A streptococcal pharyngitis [4]. Depending on genetic predisposition and the virulence of the infecting strain, 0.3-3% of people with GAS pharyngitis develop ARF [4].

Globally some 30 million people are currently thought to be affected by rheumatic heart disease, and in 2015 rheumatic heart disease was estimated to have been responsible for 305 000 deaths and 11.5 million disability-adjusted life years lost. The worst affected are the African, South-East Asia and the Western Pacific regions, accounting for 84% of all prevalent cases and 80% of all estimated deaths due to rheumatic heart disease in 2015. India has the highest global prevalence, with about 27% of all cases globally [5]. The estimated average prevalence is 0.5/1000 children in age group of 5-15 years [6]. Bhutan, in South-east Asia region, also has a high burden of cases but due to unavailability of data it's difficult to say how high. In this article, we present a case of Sydenham's chorea leading to the diagnosis of acute rheumatic fever in a female teenager in Bhutan.

CASE REPORT

A 16 years old girl presented to the outpatient department at a regional referral hospital with involuntary movements of limbs, both upper and lower for past 2 weeks duration. Left sided extremities were more affected than her right side. The symptoms had affected her daily activities and because of which she was unable to continue her school. Parents don't recall any febrile illness or any major trauma in the recent past. There was history of joints pain or swelling, rashes or chest pain. There was no history of drug intake, over the counter medications or herbal medications. There were no similar problems in the past or any family history of similar problem.

On examination, she was alert, well oriented in time, place and person. GCS E4V5M6, pulse = 82/min, regular in rhythm, normal character and volume, BP = 108/61 mmHg in right arm. There was no pallor, icterus, cyanosis, clubbing, oedema, skin rash or lymphadenopathy.

Nervous system: Higher mental function was intact, no cranial nerve deficit. Her speech was slow, reduced verbal fluency. There was jerky involuntary movement of her left sided extremities with writhing movements of her fingers. The muscle tone, deep tendon reflexes and muscle power were all symmetrical and normal. There was no cerebellar signs. Gait was unsteady with an episodic jerky movements of limbs. Cardiovascular system: No chest wall deformities, first and second heart sounds were heard normal with no murmurs or rub. Respiratory system: bilateral vesicular breaths. Abdomen: Soft with no palpable liver or spleen.

Investigations

Initial routine laboratory investigations were done. Complete blood count, renal parameters, liver enzymes and electrolytes were unremarkable (Table 2). Erythrocyte sedimentation rate was raised, 68 mm/h, Anti-streptolysin O titre = 245 IU/mL. Electrocardiogram: Normal sinus rhythm (Figure 1). Echocardiography showed mild mitral regurgitation (Video 1) with normal ejection fraction. Autoimmune panels and MRI brain not done because of its unavailability in our centre.

A diagnosis of Sydenham chorea was made based on the clinical probability and was managed with oral Phenoxymethylpenicillin 500 mg q12h and carbamazepine 200mg twice a day. Choreatic movements reduced, she was able to carry out her daily activities independently. She was discharged and currently doing well and is on follow up. She has been started on secondary prophylaxis with oral Phenoxymethylpenicillin 250

mg twice a day instead of intramuscular injection of Benzathine penicillin G as preferred by the patient and her parents.

DISCUSSION

Our case provides evidence of the persistence of acute rheumatic fever in Bhutan where clinical data and publication is very limited. In contrast to carditis and arthritis, which typically present within 21 days, the onset of Sydenham chorea usually occurs one to eight months after the inciting infection [2]. By the time patient present to medical care, they might not remember the inciting infection and therefore we cannot solely depend on history to make a diagnosis. Alternative aetiologies of acquired chorea such as autoimmune or inflammatory, cerebrovascular, drugs, infections, metabolic disorders or neoplasia should be kept in mind [2].

The diagnosis of rheumatic fever is based on the Jones criteria [Table 1]. The most common manifestation which is present in 80% of patients is arthritis, described as painful, migratory and transient. Frequently affected joints are knees and ankles. Carditis occurs in 40 - 75%, erythema marginatum and subcutaneous nodules are rare, occurring in less than 10% of patients [4]. Sydenham chorea is also a rare presentation, occurring in 10-30% [4]. In our case, the patient presented to us with Sydenham chorea which hints at a possible larger number of cases with acute rheumatic fever that may be underdiagnosed or under reported.

Carditis can be diagnosed clinically with the presence of an audible murmur consistent with aortic or mitral regurgitation on auscultation [2]. However, more recent studies on patients with acute RF have brought out the shortcomings of auscultation in identifying valve disease which does not result in hemodynamic abnormalities consisting of murmurs [4]. This has resulted in the identification of sub-clinical carditis by echocardiography [2]. In Bhutan, we have facility of echocardiography in only in three tertiary hospitals out of twenty districts in the country leading to high chances of missing the diagnosis of sub-clinical carditis.

In our case, a patient had sub-clinical carditis in the form of mitral regurgitation diagnosed with echocardiography. The patient had two major criteria, Sydenham chorea and sub-clinical carditis, and minor criteria, raised ESR and ASO titre thereby fulfilling the Jones criteria for rheumatic fever.

Rheumatic heart disease is preventable. It is a serious public health problem especially in low and middle income countries with limited capacities for the diagnosis and timely management of streptococcal infection [5]. It exerts massive economic effects globally, mainly because of premature death in children and working-age adults [7]. Globally, ARF and RHD are seen in developing nations or among disadvantaged populations within developed nations [8]. The global cost of deaths due to rheumatic heart disease in 2010 was estimated to be US\$ 2200 billion (discounted) or US\$ 5400 billion (undiscounted) [5]. The most devastating effects are on children and young adults in their most productive years because it leads to increased school absenteeism and drop-out, and lost wages [5]. The patient, in our case, she dropped out of school, but will be continuing in the next academic session after the control of the chorea.

The prevention, control and elimination of rheumatic heart disease is increasingly being recognized as an important developmental issue by the World Health Organization [5]. The barriers to prevention, control and elimination of rheumatic heart disease are poor primary and secondary prevention and access to primary health care, inadequate numbers and training of health workers at all levels, the neglect of rheumatic fever and rheumatic heart disease in national health policies and budgets, the paucity of data to enable targeting of prevention efforts, limited understanding of rheumatic fever and/or rheumatic heart disease in affected communities, and inaction on the social determinants of the disease and inequities in health [5]. In Bhutan we have communities where people prefer help from local healers than to visit health centres. In our case, the patient's father initially refused our help and wanted to go to local healer but after explaining the disease condition to him, he agreed and remained with us.

CONCLUSION

Acute rheumatic fever still continues to be a major health burden in our country. New onset chorea in childhood chorea should raise the suspicion of Sydenham's chorea. Echocardiography should be done to diagnose

sub-clinical carditis rather than depending only on clinical findings. Early diagnosis and management of acute rheumatic fever is crucial in preventing its recurrence and progressive damage to heart valves.

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Patient consent statement.

Patient consent was taken prior to the study.

Authors' contributions

TP: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Project administration; Resources; Validation; Supervision; Writing original draft; Writing review & editing.

TD: Conceptualization; Investigation, Project administration, resources, supervision, validation, visualization and writing-review and edition.

SW: Conceptualization; Investigation, validation, visualization and writing-review and edition.

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