Case Report

Atypical presentation of rheumatic fever

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ABSTRACT

Sydenham Chorea (St. Vitus dance) occurs in about 10-15% of children with acute rheumatic fever. Herein, we present the case of a 5-year-old male child with hemichorea and arthralgia. The child also presented with mild mitral regurgitation and mild aortic regurgitation. Appropriate management is essential to prevent mortality, morbidity, and psychosocial disability in such cases. We would also like to shed light on the challenges faced in the management of chorea in young children with key emphasis on the anticipation of adverse reactions to commonly used medications.

Key words: Haloperidol, Hemichorea, Jones criteria, Rheumatic fever

Acute rheumatic fever (ARF) remains a public health concern as it leads to cardiac morbidities. Sydenham chorea (SC) is seen in 10–15% of children with ARF; usually in the age group of 8–9 years with a female preponderance (2:1) [1-3]. Chorea alone can support the diagnosis of ARF according to the Revised Jones Criteria 2015 [4,5]. However, our patient presented with arthralgia and carditis. Hallmarks of SC are bilateral quasi-purpose involuntary movements, disappearing in sleep, associated with hypotonia, and emotional lability. SC usually resolves spontaneously within 3–6 months with therapy.

We hereby report a case of hemichorea in a 5-year-old male child, who subsequently developed adverse effects to the conventional treatment.

CASE REPORT

A 5-year-old boy presented with one episode of fever and joint pain involving both the knees lasting for 2–3 days which was not associated with swelling of joints and relieved on medication. The child subsequently developed involuntary movements of the right upper and lower limbs 15 days after the onset of fever and joint pain. Involuntary movements were sudden in onset, rapid, bilateral, quasi-purpose, present at rest, and absent during sleep. The child also had difficulty in speech and difficulty in walking, which coincided with the onset of involuntary movements. There was no history of sore throat, dyspnea, or effort intolerance in the child.

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On examination, the general condition of the patient was fair with the following vitals: Heart rate 104/min, respiratory rate 22/min, all peripheral pulses well felt, and blood pressure 100/60 mm Hg. Central nervous system (CNS) examination revealed positive findings in the form of dysarthria, hypotonia of the right upper and lower limbs, chorea, and abnormal gait. Signs of chorea such as Pronator sign (Fig. 1), Milkmaid Grip (Fig. 2), and Choreic hand with spooning were elicited in the child.

Laboratory findings revealed hemoglobin 12.2 g/dl, white blood cells count 8.1 × 10^3/μL, Platelets 401 × 10^3/μL, elevated erythrocyte sedimentation rate 48 mm/h, C-reactive protein 4.2 mg/L, and anti-Streptolysin-O titers 256. Throat swab culture was negative. Serum electrolytes, serum calcium, anti-nuclear antibody, thyroid function tests, and serum ceruloplasmin levels were normal.

2-dimensional echocardiography imaging of the heart revealed mild mitral regurgitation and mild aortic regurgitation suggestive of rheumatic carditis. The diagnosis of ARF was made as per the Revised Jones Criteria 2015 [5].

The patient was given a stat dose of intramuscular Benzathine Penicillin 6L IU followed by long-term penicillin prophylaxis. The patient was initiated on T. Haloperidol at 0.25 mg/kg/day for chorea and T. Aspirin at 100 mg/kg/day for carditis. The child was placed on bed rest and monitored closely for the evolution of carditis. Our patient had a rather atypical course after initiation of treatment. He developed an altered sensorium after a few days of therapy with T. Haloperidol and T. Aspirin. CNS examination was suggestive of upper motor neuron involvement, following which

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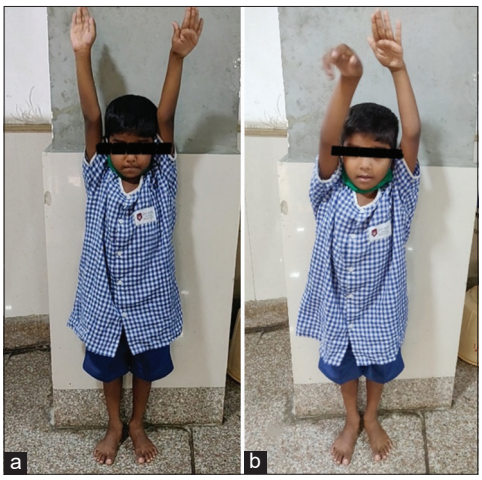

Figure 1: (a and b) Pronator sign: Arms and palms turn outward when held overhead

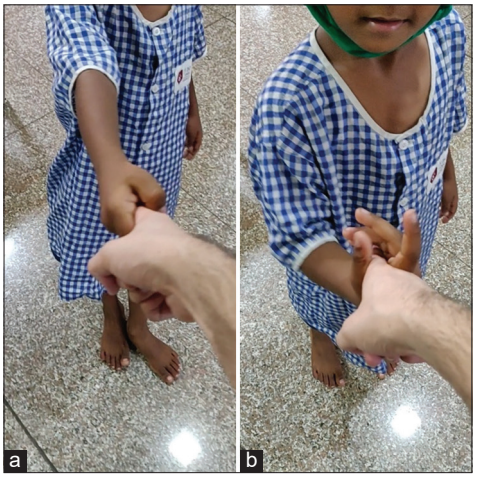

Figure 2: (a and b) Milkmaid Grip: Irregular contractions and relaxations of the muscles of the fingers while squeezing the examiner's fingers

the patient was investigated for Reye's syndrome, a complication of Aspirin ingestion in children. Since the investigations related to Reye's syndrome were normal, the dose of T. Haloperidol was revised and tapered, following which there was a significant improvement in the child's sensorium. The child is on three weekly penicillin prophylaxis and is being followed up regularly in the OPD.

DISCUSSION

ARF usually develops subsequent to an antecedent Group A Streptococcal (GAS) pharyngitis. The prevalence of ARF is high in the presence of overcrowding which facilitates the spread of GAS infections. Serotypes M types 1,3,5,6,18,29 are said to be rheumatogenic and often form highly mucoid colonies on throat culture. Incidence of both initial attacks and recurrences of ARF peaks at 5–15 years of age [1]. The pathogenesis is largely immune-mediated and based on the hypothesis of molecular mimicry.

The diagnosis of ARF is made in accordance with the Jones Criteria, revised in 2015 (Table 1) [5,6]. The unique features in our patient were age, male gender, and chorea affecting one side of the body associated with arthralgia, and carditis. Our case belonged to the moderate/high-risk population.

A case of SC reported from Saudi Arabia had features analogous to our case wherein a 5-year-old male child presented with choreiform movements and a history of frequent episodes of pharyngitis in the past. The case, however, does not discuss the management of SC in young pre-school children [7]. Another report from a low-prevalence European setting highlights the importance of keeping a high index of suspicion for ARF in patients presenting with choreiform movements even in settings where the incidence of ARF is low. It also emphasizes the need to rule out subclinical carditis in patients without any signs and symptoms of carditis [8].

SC occurs in 10–15% of patients with ARF and usually manifests 1–6 months after the episode of streptococcal pharyngitis. Emotional lability, incoordination, poor scholastic performance, uncontrollable quasi-purposive movements, and facial grimacing are characteristics of the disease. These features disappear during sleep and are exacerbated by stress. All patients with ARF should be placed on bed rest, more so in our case who was diagnosed with subclinical carditis.

Once diagnosed as ARF, the patient should receive 10 days of oral penicillin/amoxycillin or a single IM injection of benzathine penicillin (after test dose) to eradicate GAS, as in our case, followed by long term penicillin prophylaxis [9,10]. Since our patient had ARF with carditis but without residual heart disease, he was started on 3 weekly penicillin prophylaxis to be continued till 21 years of age as secondary prevention. Aspirin in the dose of 100 mg/kg/day in 4–5 divided doses for 2–3 weeks followed by 60–70 mg/kg/day is recommended in patients with carditis without cardiomegaly or congestive cardiac failure [11,12]. Haloperidol 0.25–0.5 mg/kg/day is recommended for the management of chorea [12]. Adverse reactions such as dystonia, Parkinsonism, sleepiness, and forgetfulness should be anticipated, and the dose should be revised accordingly [13]. Alternative approaches with valproate, carbamazepine, and benzodiazepines should be considered. Treatment with high-dose prednisone is thought to have faster remission rates.

SC usually resolves spontaneously within 3–6 months, rarely lasting longer than 1 year. The involuntary movements in our patient subsided over a period of 4 weeks following which Haloperidol was discontinued. Improvement in the child's handwriting (Fig. 3) could be considered a reliable parameter of recovery. Awareness regarding the side effects of medications used to treat SC is vital.

CONCLUSION

Awareness and sensitization of rheumatic chorea should be thought of when a child presents with repetitive, quasi-purposive

Table 1: Revised jones criteria 2015

Criteria	Low-risk population	Moderate/high-risk population
	ARF incidence ≤2 per 100000 school-aged children or all-age RHD prevalence of 1 per 1000 population year	Children not clearly from a low-risk population
Major criteria		
1. Carditis	Clinical and/or subclinical	Clinical and/or subclinical
2. Arthritis	Polyarthritis	Monoarthritis, polyarthritis, and/or polyarthralgia
3. Chorea	Chorea	Chorea
4. Erythema marginatum	Erythema marginatum	Erythema marginatum
5. Subcutaneous nodules	Subcutaneous nodules	Subcutaneous nodules
Minor criteria		
1. Carditis	Prolonged PR interval	Prolonged PR interval
2. Arthralgia	Polyarthralgia	Monoarthralgia
3. Fever	≥38.5°C	≥38°C
4. Inflammatory markers	Peak erythrocyte sedimentation rate ≥60 mm in 1hr and/or C-reactive protein ≥3.0 mg/dL	Peak erythrocyte sedimentation rate ≥30 mm in 1hr and/or C-reactive protein ≥3.0 mg/dL

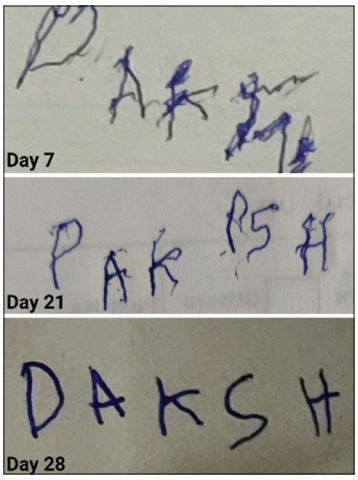

Figure 3: Gradual improvement in handwriting of the patient

involuntary movements which disappear in sleep. However, the presentation of arthralgia demonstrated the atypicality. Diligent history taking with special emphasis on grimacing, changes in handwriting, unable to button the shirt should be highlighted. Pronator sign, milkmaid sign, and straight walking sign should be performed. Treatment should be meticulous with three weekly penicillin prophylaxis and aspirin in case of cardiac involvement. Close diurnal monitoring of sleeping pulse rate, peripheral pulses, handwriting, and daily activities are to be recorded and monitored with a watchful eye for signs of failure.

AUTHOR CONTRIBUTION

Concept and design of study or acquisition of data or analysis and interpretation of data: Fehmida Najmuddin, Sushil Yewale, Keya Lahiri.

Drafting the article or revising it critically for important intellectual content: Keya Lahiri, Sushil Yewale.

Final approval of the version to be published: Keya Lahiri, Fehmida Najmuddin, Anand Sude.

Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved (guarantor): Fehmida Najmuddin, Keya Lahiri.

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