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Case Report

An atypical case of Rheumatic Chorea in a rural tertiary health centre of South India

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ABSTRACT

Varied presentation of infectious diseases like the rheumatic fever is on the rise especially in rural areas where a great proportion of communicable diseases are undiagnosed and untreated. Keen clinical examination and early treatment with minimal laboratory aid reduce healthcare expenditure in low socioeconomic regions. An 18-year-old adolescent girl presented with acute onset of motor tic for 2 weeks which progressed to vocal tic in a span of 6 weeks. As per the initial presentation, the patient was worked up as a case of Transient Tic Disorder and later as Tourette Syndrome. All the test results including Rheumatic Heart Disease workup were negative. But the patient developed Pure Chorea 2 weeks after this. The patient was treated on a presumptive diagnosis of Rheumatic Chorea and is now in remission for 6 months. Even though tics and other movement disorders can be associated with Rheumatic chorea, it is seldom to present as pure tic disorder at the first encounter and in relatively uncommon age group making this case special. Rather than early referral to movement disorder center, detailed observation of disease progression can prevent unnecessary work up.

Keywords: Rheumatic Chorea, Transient Tic disorder, Tourette syndrome

INTRODUCTION

Rheumatic Chorea is a neurological manifestation of Rheumatic Fever (RF) occurring in 5.4% of patients with RF in South India as indicated by Vaishnava et al.[1] The most common presentation being Rheumatic Carditis amounting to 97%.[2] Rheumatic Chorea is a major criterion for the diagnosis of acute rheumatic fever and according to the modified Jones' criteria, it's presence alone is sufficient to make this diagnosis mostly in developing countries.[2] The typical age of onset is 5-14 year. The first episode of chorea tends to be 'pure chorea', but this varies in different communities.[3,4] Evidence of recent streptococcal infection as shown by a raised antistreptolysin titer or an anti-DNase titer varies in different regions but was demonstrated in studies from Turkey and Australia.[4,5]

CASE REPORT

An 18-year-old adolescent girl presented with the acute onset of eye blinking, shoulder shrugging, mouth opening, facial grimacing and lip-smacking for 2 weeks. She stressed out when restrained from performing the activity and relieved on performing the same. The involuntary movements ceased at night and also on distraction. The involuntary rapid, brief, non-rhythmic movements were categorized as simple and complex motor tic involving mainly the face and shoulder. A provisional diagnosis of Transient Tic disorder was made. Work up was done to rule out secondary causes for it. As the day progressed she developed vocal tic consisting of sniffing, snorting and frequent throat clearance 6 weeks after the motor tics. The diagnosis of early stages of Tourette syndrome was considered as both motor and

vocal tics were present together. Even though the tics were just 6 weeks old at the time of presentation.

At the time of presentation, there was no fever, fatigability, malaise, weight loss, loss of appetite, previous seizure episode, skin lesions or hypersensitivity, joint pain, chest pain, pain abdomen, bowel and bladder disturbances, menstrual irregularities, mood and cognitive disorder, or allergic symptoms. We couldn't elicit any positive history in terms of Obsessive Compulsive disorder (OCD), Attention Deficit Hyperactivity Disorder (ADHD), learning disabilities, the utterance of obscene words, persistent drug usage or any other psychiatric illness and similar complaints among family members from the patient and her father. On thorough probing, an episode of 2-day fever without cold, cough or a sore throat was obtained 6 weeks before the onset of Motor tic which resolved spontaneously with symptomatic treatment. She had never been hospitalized or consulted a physician for any illnesses apart from this in the past 1 year.

On neurological examination, no neurocutaneous markers were present. No signs of meningeal irritation were present. The speech was normal except for occasional stuttering. Subjective and objective mood assessment revealed euthymic state except she felt unhappy when people scorn her condition and felt anxious when restrained from performing the movements. Higher mental functions like perception, cognition, and judgment were good. Insight was present. Examination of cranial nerves, sensory and, the motor system was normal. Tests for Cerebellar functions were within normal limits.

Initially as the patient presented with tics, work up was done to rule out secondary causes. All the investigations like complete blood count, electrolytes, acute phase reactants, Liver Function Tests (LFT), Renal Function Tests (RFT) and thyroid profile were within normal limit ruling out infectious etiology, metabolic, endocrinal causes respectively.

Autoimmune diseases like Systemic Lupus Erythematosus (SLE) and Anti Phospholipid Antibody (APA) syndrome were excluded by negative ANA (0.3), nil thrombotic abnormalities and didn't satisfy any criteria for SLE.

Post-infectious causes like Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections (PANDAS) were out of the picture as Total Leukocyte count (TC): 4600, ESR: 12 mm/hr, C-Reactive Protein (CRP) <0.6 and C-Reactive Protein (DC) were within normal limit and ASLO was negative and no association could be traced with reference to a probable viral infection and the symptoms. Serum Ceruloplasmin was 26.80 mg/dl (normal 20-35 mg/dl) and Ophthalmic examination by a slit lamp did not reveal any Kayser Fleischer rings eliminating Wilson's disease.

No growth was observed from the throat swab. ECG was normal with no wave or interval abnormality like PR prolongation. Echocardiogram was normal with normal chamber volumes, good LV function, EF: 62% and normal valves. No regional wall motion abnormality was detected. No clots or pericardial effusion was present precluding rheumatic carditis. ASLO <200 IU/ml at the first visit and no raise was recorded on further visits. Anti-DNase B was also negative (<85 IU/ml). No abnormality was detected on MRI eliminating CNS tumors. EEG was normal excluding any subtle seizure disorders. All the possible causes were eliminated. Tics were treated with 0.5 mg of Haloperidol (0.025 mg/kg/day) Bd. But the patient reported worsening of symptoms.

On subsequent visits, her gait became faintly clumsier with supplementary hand and leg swings in addition to the tics. It progressed to typical chorea in a span of 2 weeks. She also noticed a significant variation in her penmanship in comparison to her premorbid self. As a result, she failed to materialize for her exams. She also had disturbed sleep. She was emotionally disturbed and easily lost her temper. A clinical diagnosis of rheumatic Chorea was made. Development of chorea ruled out our provisional diagnosis of Transient tic disorder and Tourette syndrome. Haloperidol was replaced with 200mg of Carbamazepine (15-20 mg/kg/day) OD which was slowly titrated up to 400mg in two divided doses. 1.2 million units of Benzyl penicillin via deep intramuscular route was started and repeated every 28 days.[6,7] The patient experienced significant improvement in her symptoms over 2 weeks. The patient is now in remission for past 6 months. She resumed her studies as before.

DISCUSSION

The common age group for Rheumatic Chorea is 5-14 years with first episode occurring as early as 10.4 years.[4] In developing countries like India especially rural areas, the first episode of Chorea can occur as late as 18 years.

Tics are usually the associated symptom present along with Chorea at later stages. But Tics being the only presenting complaint diverting our differentials towards Transient Tic Disorder or Tourette syndrome is seldom. No reports are available as to what percentage of cases of Rheumatic Fever are present in association with other involuntary movements and in early stages of the disease. Pure Chorea is present in only 5.4% of cases in South India.[1] Diagnostic dilemma is one of the major challenges in the initial stages of the disease.

The patient's uncertain history of 2-day fever cannot easily be categorized under Group A Beta-Hemolytic Streptococcal (GABHS) infection as it was not associated with a sore throat or skin infection. It resolved spontaneously by symptomatic treatment without any antibiotics suggesting viral etiology. ASLO and Anti Dnase titers drawn just after 8 weeks of this febrile

episode didn't show any raise. In young adults, the initial titers of ASLO and Anti DNase B are low compared to older children. By the time chorea presents, the streptococcal antibody titers are often considered to be low.[8] So in presumptive treatment of any form of movement disorders like Tics, a differential diagnosis of Rheumatic Chorea should be kept in mind.

Even without a history of antecedent GABHS infection and low ASLO/Anti DNase B titers, Rheumatic Chorea cannot be foreclosed. A trial of treatment with antibiotics is beneficial.

Haloperidol is an economical drug used in developing countries but has comparatively greater side effects than Carbamazepine, Valproic acid and other drugs are also used in Chorea. Her symptoms aggravated with Haloperidol due to unknown reasons. One possible explanation is the inability to start on high dose Haloperidol due to patient noncompliance and not treating the primary disease with Penicillin which was unrevealed at that time. Carbamazepine is usually well tolerated except for allergies. There are no specific guidelines for the symptomatic treatment of Chorea with different agents. It mostly depends on clinical experience based on the patient response in different cases.[9,10]

Keen observation of the clinical progress of a patient with movement disorder decreases unnecessary investigations like MRI, EEG, and referral to higher movement disorder center at initial stages. Various presentation of familiar diseases like GABHS is more prevailing in rural areas than other uncommon secondary causes for any movement disorders.

CONCLUSION

The first episode of Pure chorea can occur as late as 18 years in developing countries. Rheumatic Chorea to be considered as a differential diagnosis in any forms of movement disorders occurring in rural areas of developing countries.

Keen observation on the progression of symptoms leads to diagnosis rather than premature referral to higher movement disorder centres and unnecessary workup.

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