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Case report

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ABSTRACT

Sydenham chorea (SC) is the most common acquired cause of chorea in children. SC is the neurological manifestation of rheumatic fever. Rheumatic fever is still reported in Saudi Arabia, although less frequently. According to modified Jones criteria, carditis and arthritis are the major manifestations of acute rheumatic fever (ARF) in children.

SC is often seen in isolated form; however, it can occur in association with other clinical manifestations of ARF like carditis. It has been reported that silent, mild valvular regurgitation has been found in patients with pure chorea.

SC is characterized by abnormal body movement, associated with behavioral changes. It is an autoimmune disease that occurs following an infection with Group A beta-haemolytic streptococcal infection that is prevalent in Saudi Arabia.

Despite recent advances in child health care services, RF continues to occur in Saudi Arabia. We describe a 12-year-old Saudi girl with SC and clinically inaudible, but echocardiographically significant mild valvular regurgitation.

The diagnosis of Sydenham chorea should be considered in young children with choreiform movements. Doppler echocardiography may be useful in detecting silent valvular regurgitation and in deciding the duration of penicillin prophylaxis.

1. Introduction

Sydenham chorea (SC) is a neurological disorder of childhood that occurs after infection with Group A beta-hemolytic streptococcus (GABHS) [1]. It may be the sole manifestation of rheumatic fever (RF). The prevalence of SC in developed countries is rare However, the disease is still found in developing countries [2].

2. Case report

A 12-year-old Saudi girl was brought to the pediatric ER for worsening involuntary abnormal movements of the face and extremities, clumsiness, and abnormal speech. The symptoms began 3 weeks before presentation. The symptoms gradually worsened to include dropping eating utensils, facial grimacing, random tongue movements, and slurred speech. The symptoms stopped during sleep and become aggravated with stress. The child had no past medical history of significance except repeated attacks of tonsillitis. History of a mild throat infection that resolved spontaneously several weeks before the onset of abnormal movement. She had no history of drug intake or allergies. No family history of behavioral or neuropsychiatric disorders.

She was vitally stable and afebrile. The ear-nose and throat examination revealed hypertrophic non-inflamed tonsils. Examination of the chest and abdomen were normal. Examination of the heart revealed normal heart sounds and no audible murmurs. Examination of the musculoskeletal system was normal. Skin examination was normal. Neurologically, she was conscious, alert and oriented to time, person, and place. She was cooperative and intelligent. She showed choreiform movements of the face, upper, and lower extremities. Cranial nerves were intact, and her sensation was normal. Motor system examination
revealed normal muscle tone and strength globally. She had abnormal hand grip with difficult maintaining a tetanic contraction with her hands. “Milkmaid’s sign”. She showed sporadic tongue protrusion (darting tongue). Deep tendon reflexes were normal and symmetrical on both sides, and her toes were down going. She ambulated with assistance to avoid falling down. No other manifestation of RF was detected.

Routine laboratory studies were normal. The erythrocyte sedimentation rate (ESR) was 18 mm/h and C-reactive protein (CRP) was negative. A throat culture showed negative results and anti-streptolysin O titer (ASOT) was unremarkable.

Antinuclear antibody (ANA), anti-DNA and complement 3 and 4 levels were normal.

Thyroid function tests were normal.

Brain imaging (CT and MRI) and electroencephalogram (EEG) were normal.

Color Doppler echocardiographic evaluation revealed mild but significant mitral regurgitation. The case was diagnosed as SC. Treatment was given to her in the form of monthly long acting benzathine penicillin (1.2 million units administered intramuscularly) as a long-term prophylaxis for at least 10 years and carbamazepine (200 mg administered orally 3 times a day). She showed good control of her choreiform movements. During follow-up visits, she continued to improve and was back to her baseline state by 6-month follow-up appointment.

3. Discussion

The disease presents abruptly with neurological and psychological symptoms up to 6 months after a streptococcal pharyngitis. The muscle weakness of SC presents as an inability to do a continuous contraction, known as a "milkmaid’s grip," in which patients relax and tighten their fists intermittently when asked to grip the examiner’s fingers. The choreiform movement leads to disturbance of gait, dropping of objects, and dysarthric speech [1].

Our case showed negative ASOT. Although elevated levels of ASOT can be found in patients with SC, 20% of patients may not have these findings. So, that their absence does not exclude the diagnosis of SC. Diagnosis of SC is mainly clinical after exclusion of other causes of chorea.

Our case showed normal routine laboratory studies, ESR, CRP, thyroid function tests ANA, anti-DNA and complement 3 and 4 levels.

Also, Brain imaging (CT and MRI) and EEG were normal. These investigations help to differentiate this case from other conditions like collagen disease, encephalitis, drug intoxication, familial chorea (eg, Huntington disease), atypical seizures, stroke, Tourette syndrome, thyroid or parathyroid abnormalities, Lyme disease, or Wilson disease [4].

Recently, color Doppler echocardiography has disclosed subclinical valvular regurgitation in some patients with ARF manifested as isolated arthritis or pure chorea [5].

As diagnostic criteria for sub-clinical carditis are lacking, the 2015 Jones Criteria suggest that the WHF criteria may be used to distinguish physiological from pathological regurgitation. Both clinical and sub-clinical carditis is considered a major manifestation of ARF in both low- and high-risk populations [5].

Our patient showed good response to Carbamazepine therapy without reported side effects. Carbamazepine was reported to be effective for cases of childhood nonhereditary chorea [3].

4. Conclusion

Childhood chorea of new onset should raise the suspicion of SC. In suspected cases of SC, a careful cardiac examination and color Doppler echocardiogram must be performed.

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Declaration of Competing Interest

None to be declared.

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