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Sydenham Chorea in a Girl with Dextrocardia and Situs Inversus

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ABSTRACT

Sydenham Chorea (SC) is a childhood disease that most frequently occurs after infection with group A β-hemolytic streptococcus species. SC is the most commonly acquired childhood chorea, usually affecting 5 to 15-year-old children. Dextrocardia situs inversus refers to the heart being situated on the right side of the body. We found a case of SC with coexisting dextrocardia and situs inversus, which is rare to our knowledge. A 14-year-old girl was referred to Ali-Ibn-Abiltaleb Hospital due to choreoathetotic movements more in her upper than lower extremities, which impaired her usual function. Antistreptolysin O (ASO) titer was high. Her throat culture was positive for streptococci beta hemolytic. In cardiac study, moderate mitral regurgitation, tricuspid regurgitation, diastolic dysfunction, conus in the Right Ventricle Outlet (RVOT), right-sided aortic arch, dextrocardia, and situs inversus were reported. This was a rare case with SC that had such anomalies.

1. Introduction

Sydenham Chorea (SC) is a childhood disease that most frequently occurs after infection with group A β-hemolytic streptococcus species (GAS) and Acute Rheumatic Fever (ARF). SC is in fact a complication of ARF. SC is the most commonly acquired childhood chorea, usually affecting 5 to 15-year-old children (1).

Chorea is observed in 18 - 36% of all patients with ARF. SC could remain latent and present up to 6 months following an infection with this class of organism (1, 2). Most patients are presented with non-neurological features of ARF, such as carditis and arthritis (3).

This disorder is an antineuronal antibody-mediated neuropsychiatric disorder caused by a post- streptococcal autoimmune condition affecting control of movement, mood, behavior, and potentially the heart. Anti-D1R and anti-D2R autoantibodies were significantly higher in patients with SC compared to controls. In addition, anti-D2R titers were correlated to antistreptolysin-O titers (4). Moreover, up to 60% of individuals who present with SC will develop rheumatic heart disease.

SC is the most common form of acquired chorea and its clinical presentations include involuntary choreatic movements, voluntary movement incoordination, muscular weakness, and hypotonia. Its accompanying psychiatric disorders also include emotional lability, hyperactivity, distractibility, obsessions, and compulsions. SC has been associated with increased frequency of psychiatric disorders (5).

Although the motor features of SC change to spontaneous remission in the majority of patients, persistent chorea is observed in a significant proportion of patients (3). Cardiac involvement has been reported to occur in 30 - 70% of patients with their first attack of RF. Pancarditis is the most serious and the second most common complication of RF (50%). Murmurs of ARF are caused by valve regurgitation, while those of chronic RF are produced by valve stenosis. A long latency period exists between streptococcal pharyngitis (1 - 6 months) and the onset of chorea. Therefore, a history of sore throat is not frequently obtained. Indeed, patients with chorea do not often demonstrate Jones criteria, which include the "major criteria" for diagnosis, such as carditis (50 - 70%) and arthritis (35% - 66%). These are followed in frequency by chorea (10 - 30%) that has been demonstrated to have a female predominance, subcutaneous

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nodules (0 - 10%), and erythema marginatum (6). The minor criteria include polyarthralgia or monoarthralgia in high-risk populations, fever (≥ 38.5 or ≥ 38 °C) in high-risk populations, Erythrocyte Sedimentation Rate (ESR) ≥ 60 mm in the first hour or ≥ 30 mm in high-risk populations, C-Reactive Protein (CRP) ≥ 3.0 mg/dL, and prolonged PR interval after accounting for age variability (unless carditis is a major criterion). However, heart anomalies were not reported in many cases of ARF carditis.

The Jones criteria have been modified throughout the years, most recently in 1992, to aid clinicians in diagnosis of initial attacks of ARF and to minimize overdiagnosis of the disease (7). Recently, these criteria have been changed minimally according to low-risk and high-risk populations (8).

Dextrocardia situs inversus refers to the heart being a mirror image and situated on the right side of the body. Additionally, dextrocardia situs inversus totalis refers to all visceral organs being mirrored. On the other hand, isolated dextrocardia refers to non-displacement of other organs. Situs inversus is a rare abnormality typically posing a diagnostic dilemma during routine evaluation of acute abdominal emergencies, such as acute appendicitis and cholecystitis (9).

Although patients with dextrocardia situs inversus do not present with any medical problems due to this disorder, they may be prone to a number of bowel, esophageal, bronchial, and cardiovascular disorders, such as double outlet right ventricle, endocardial cushion defect, and pulmonary stenosis (10). Although the incidence of SC has diminished because of aggressive antibiotic treatment, both isolated cases and epidemics persist, necessitating emergency physicians to be familiar with this disease (1). We found a case of SC with coexisting dextrocardia and situs inversus totalis, which is a rare case.

2. Case Presentation

A 14-year-old girl was referred to Ali-Ibn-Abiltaleb Hospital due to choreathetotic movements more in her upper than lower extremities. Choreathetotic movements impaired her normal function and prevented her from going to school for 4 days. She had a history of skin rashes, including pink and red macules (flat spots) and papules (small lumps) on the abdomen before the chorea attack. The rash gradually spread out and as it did so, the skin in the center of lesions changed to normal, thus forming the typical spreading marginate or annular eruption. She was unable to walk and do her homework. Dysarthria and grimacing were observed in her examination. In addition, mild gallop was heard during cardiac examination. Moreover, she had presented emotional lability and hyperactivity in the recent days. She had a history of upper respiratory tract infection in the previous 4-6 weeks that was not appropriately treated. There were no symptoms or signs of arthritis in her clinical examination. However, her throat culture examination was positive for group A streptococci beta hemolytic. Moreover, her Anti-Streptolysin O (ASO) titer was 3200, White Blood Cells (WBC): 6500, Red Blood Cells (RBC): 5.81, Hemoglobin (Hb): 11, Hematocrit (Hct): 33.7, Mean Corpuscular Volume (MCV): 58, Mean Corpuscular Hemoglobin (MCH): 18.9, Mean Corpuscular Hemoglobin Concentration (MCHC): 32.9, platelet: 374000, ESR: 27, CRP: +2, urea: 20, Creatinine (Cr): 0.8, sodium (Na): 145, potassium (K): 4.5, Serum Glutamic-Oxaloacetic Transaminase (SGOT): 22, Serum Glutamic Pyruvic Transaminase (SGPT): 21, Alkaline Phosphatase (ALK PH): 375, bilirubin (Bil total): 0.6, bilirubin (Bil direct): 0.2, T3: 0.8, T4: 11.1, Thyroid Stimulating Hormone (TSH): 0.4, and Rheumatoid Factor (RF) was negative.

Chest X-ray showed dextrocardia and situs inversus (Figure 1). Besides, echocardiography revealed moderate mitral regurgitation (Grade II), tricupid regurgitation (Grade II), Ejection Fraction (EF) of 55%, Left Ventricular End-Diastolic (LVED) pressure of 3.5 mmHg, Left Ventricular End-Systolic (LVES) pressure of 2.1 mmHg, normal Left Ventricular (LV) size, diastolic dysfunction, conus in the Right Ventricle Outlet (RVOT), right-sided aortic arch, dextrocardia, and situs inversus (Figures 2 and 3) (Video 1) [To watch the video, please refer to the html format]. She refused MRI performance. Therefore, brain Computed Tomography (CT) scan was performed the result of which was normal. Therapy with antifailure and anti-inflammatory medications was started for her carditis. Additionally, valproate, haloperidol, and biperiden were used for treatment of chorea. The symptoms of chorea subsided after 3 days. After that, she was discharged from hospital.

3. Discussion

Evaluation of chorea in pediatric patients should include testing for Group A Streptococcal (GAS) infection through throat culture and ASO titers (11).

Despite improvements in socioeconomic status and standards of healthcare services, RF continues to occur in Saudi Arabia and other countries although with decreasing frequency. This disease is most commonly observed in school-aged children, but it can also occur in younger age groups. Carditis and arthritis are the major clinical

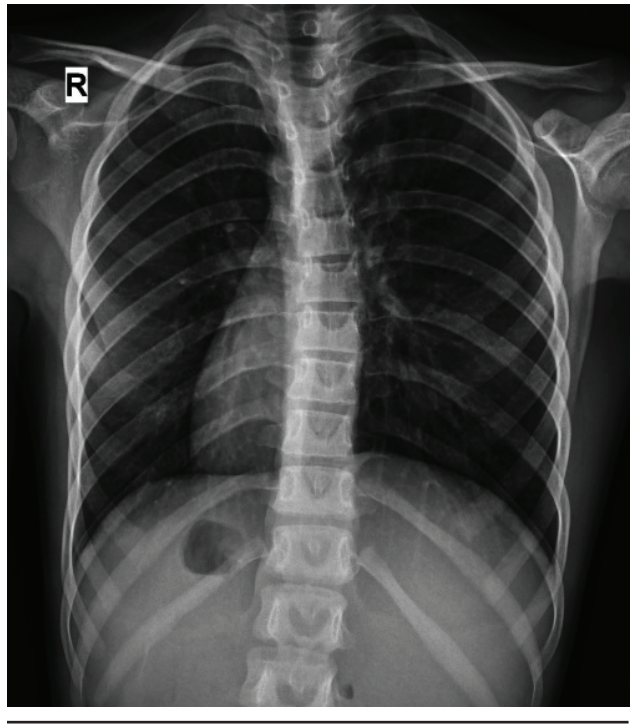

Figure 1. Dextrocardia and Situs Inversus Totalis

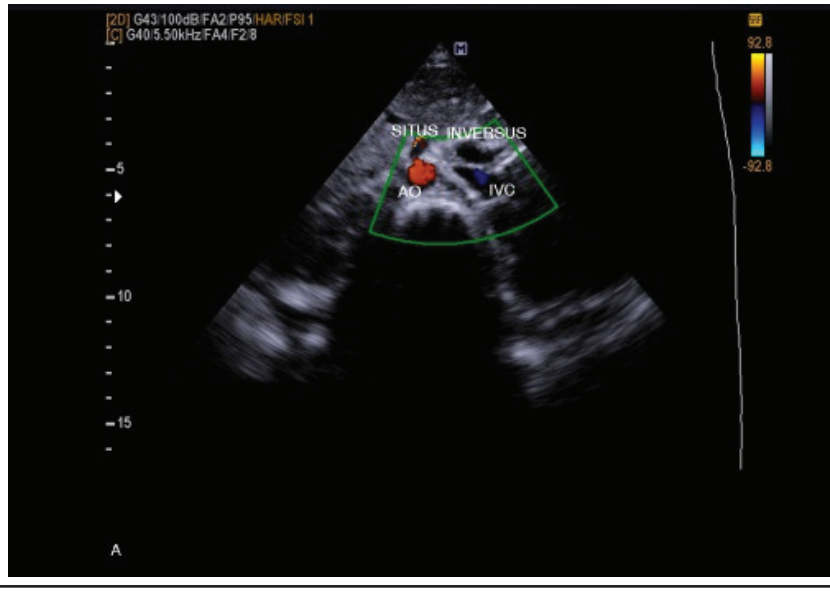

Figure 2. Dextrocardia and Situs Inversus, Right Aortic Arch

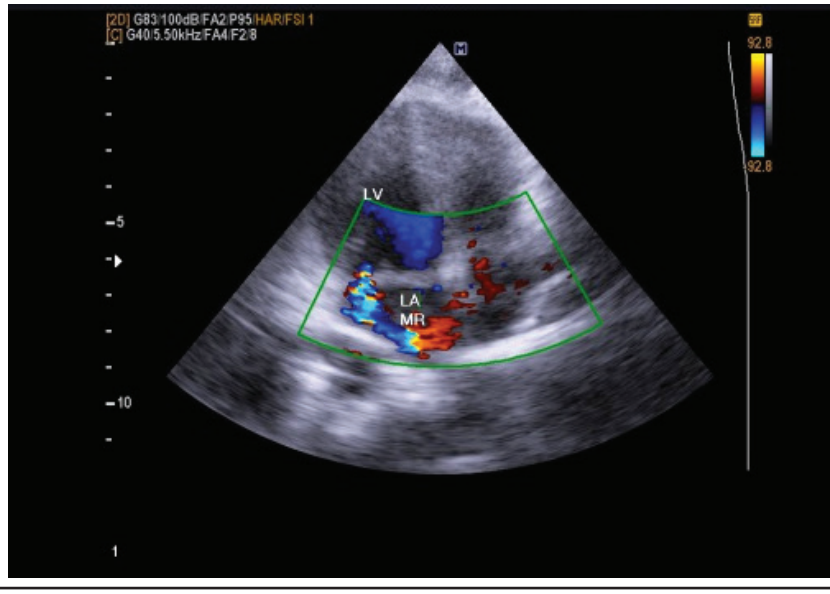

Figure 3. Mitral Regurgitation

symptoms on presentation of ARF in young children. However, rheumatic chorea is infrequently reported in young children. A case of SC in a 5-year-old boy was reported by Lardhi (12). Despite being rare, diagnosis of SC should always be considered in young children with choreiform movements (12).

It is believed that younger children are more likely to have carditis and arthritis and less likely to have chorea. A case of rheumatic chorea in a 3 and 1/3-year-old boy was reported by Ramanan et al. The patient was treated with penicillin, prednisolone, and sodium valproate and responded favorably (13).

Zomorrodi and Wald conducted a retrospective chart review to determine the causes of childhood chorea seen in a large pediatric hospital in an area endemic for ARF (14). The causes included SC (n = 79), postoperative cerebral ischemia (n = 1), and basal ganglion infarct (n = 2). In addition, 76 children (71%) with SC were female (14). Also, 96% of children who had acute chorea suffered from SC. As in our case, the most common, concurrent, major Jones criterion in the study by Ramanan et al. was carditis (13). Arthritis, erythema marginatum, and subcutaneous nodules were uncommon in this population (13). In our study, erythema marginatum occurred before chorea. Cavusoglu et al. described a 19-year-old boy with SC, rheumatic heart disease, and cardiomegaly (15). He was also found to have non-compaction of the ventricular myocardium (NVM), which has recently been categorized as primary genetic cardiomyopathy (15). The disease might become clinically overt during childhood and adolescence, but symptoms onset may be delayed until the 7th decade of life.

Neuromuscular disorders are the most frequent non-cardiac abnormalities associated with NVM. In the current report, a noticeable problem was the poor health of the patient's family and absence of persistent control for her disease. If her sore throat had been treated primarily, this problem would not occur. Indeed, she did not appropriately follow the treatment for her chorea and carditis.

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Authors' Contribution

All authors contributed to all stages of the case report.

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The authors declare that there is no conflict of interests.

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