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CASE REPORT

The Diagnosis of Rheumatic Fever by the clinical presentation of Sydenham Chorea: Case report

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Abstract

Sydenhams chorea is characterized by involuntary movements of the distal limbs and face region and mainly affects females during their childhood. It occurs by complication of pharyngotonsillitis caused by beta hemolytic streptococcus of the A group. This case report aims to describe a case of rheumatic fever diagnosed from a clinical picture of Sydenhams chorea. A descriptive case study of a female child was carried out, she was twelve years old, she sought the emergency care unit with involuntary movements in lower and upper rights limbs and face for five days, she had a previous history of infection an upper respiratory infection with self-limited evolution. The physical examination found, was in good general condition, with extension and flexion limbs movements, opening and closing hands movements, finger and face movements, complementary exams, they did not demonstrate any alterations, except ASLO of 217IU/ml. It was started a treatment with haloperidol, benzathine penicillin and and school progress evolved asymptomatic with improved choreatotic movements. This report demonstrates the relation between Sydenhams chorea and the late diagnosis of rheumatic fever, in addition to explaining the good prognosis from adequate and early therapeutic management.

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INTRODUCTION

Sydenham's chorea (CS) is characterized by involuntary, sudden, rapid, arrhythmic and irregular movements, predominantly in the distal region of the limbs and face, occurring mainly during childhood and in females. In general, it is an extemporaneous presentation of Rheumatic Fever (RF), triggered by a complication of pharyngotonsillitis caused by group A beta-hemolytic streptococcus (Streptococcus pyogenes). This report describes a case of Rheumatic Fever, diagnosed from the clinical picture of Sydenham's Korea.

CASE REPORT

MCT, 12 years old, female, sought care in the emergency department, accompanied by her mother due to involuntary movements in the upper and lower limbs (RUL and MID) and face that started five days ago. The mother reported that her daughter had presented with an upper airway infection for approximately 15 days, with a self-limiting evolution. It also reported previous, recurrent cases of upper airway infection in childhood and attention deficit hyperactivity disorder. On physical examination, she was in good general condition, active, collaborative, and lucid. Irregular heart rhythm with a rate of 108 bpm and absence of murmurs. On neurological examination, he had involuntary movements of extension and flexion of the MSD and MID, opening and closing of the hands, finger movements and grimacing that worsened when occluding the eyes. Painful and tactile sensitivity was preserved and Babinski's sign was negative. The gait showed an increase in the support base with a slight movement of launching the right foot, without imbalance, negative Romberg. He also presented dysmetria on the right and fractionation of the MSD movement in the index-nose test. Preserved eye movement and isochoric and photoreactive pupils. Haloperidol 2.5mg was started, increasing by 0.5mg every 3 days until reaching the maximum daily dose of 5mg, to control the movement disorder and the etiological investigation was initiated. Laboratory tests performed 10 days after the onset of the condition showed ASLO: 217 IU/ml (RV: 214 IU/ml), C3: 91.50 mg/dL, C4: 16.90 mg/dL, FAN: Reagent 1/ 160 and negative Anti-SM. There were no alterations in the cardiological assessments, Echocardiogram and Electrocardiogram, as well as in the cranial tomography. Despite the improvement in the movement pattern after starting Haloepridol, treatment with Benzathine Penicillin, 1,200,000IU every 21 days, was proposed. After one month, due to remission of choreoathetotic movements, the use of Haloperidol was suspended and Benzathine Penicillin was maintained. The patient remains asymptomatic, fulfilling the therapeutic proposal of prophylaxis, without intercurrences so far.

DISCUSSION

Rheumatic fever (RF) is a late complication of pharyngotonsillitis caused by Lancefield group A beta-hemolytic streptococcus or Streptococcus pyogenes, which occurs primarily in underdeveloped countries with poor hygiene conditions. It is estimated that about 0.3% to 3.0% of individuals infected with strains of Streptococcus known to be rheumatogenic will actually develop RF[1,2]. Patients may develop carditis (50–78%), arthritis (35–88%), chorea (2–19%), erythema marginatum (<6%), and subcutaneous nodules (<1–13%)[3]. In Brazil, of the 10 million annual cases of pharyngotonsillitis, 30,000 are diagnosed with RF, and 5 to 36% are diagnosed with CS[4].

Sydenham's chorea was described by Thomas Sydenham in 1686 and has been associated with rheumatic fever since 1956.[5] It is an important neurological manifestation, according to the last modification of the Jones criteria in 2015[6], a sufficient criterion by itself for the diagnosis. of the disease[7,8]. Chorea usually develops 4-8 weeks after the onset of pharyngotonsillitis, unlike carditis and arthritis, which have a more recent onset between 2 and 3 weeks after infection[7]. Data on the incidence of Sydenham's chorea in Brazil are scarce in the literature, but the frequency of this pathology, according to a retrospective analysis by the University of São Paulo, is similar to epidemic episodes in developed countries[8]. In a study carried out in 7 centers in the State of São Paulo (Brazil) with 786 patients, chorea was present in 38% of patients with rheumatic fever (299 patients)[8], the peak incidence occurs between 5 and 15 years of age, gender being female the most affected[7].

The outbreak of chorea is late, insidious and nonspecific. The patient may have emotional lability and muscle weakness that later progress to rapid, involuntary, uncoordinated movements of the limbs and face, which worsen with adrenergic discharge and tend to disappear during sleep. They may also present dysarthria, difficulty in writing, obsessive compulsive disorder, anxiety, aggression, irritability, personality changes. It usually lasts an average of two to three months, but can last for more than a year[4].

The pathophysiology of the disease is multifactorial and today, with population studies, it was possible to observe that there is a pattern of susceptibility to rheumatic fever. The disease is considered autoimmune and starts from a cross reaction, where antibodies attack the tissue itself due to the cellular mimicry made by the rheumatogenic strains. From then on, the individual's immune response pattern directs the clinical picture, in the case of a Th1 cellular response, the clinical picture is carditis, while the Th2 humoral response evolves mainly with chorea and arthritis[4].

The diagnosis of the first outbreak is based on the Jones criteria, confirmed by the presence of two major criteria or one major and two minor criteria, associated with previous streptococcal infection. The use of 3 minor criteria can be used for the diagnosis of recurrent or relapsed RF. (Table 1)[9,10].

Proven evidence of previous infection with group-A B-hemolytic Streptococcus (positive oropharyngeal culture, positivity in rapid tests for the detection of streptococcal antigens, high titers of antistreptococcal antibodies).

Treatment consists of controlling the inflammatory process and eradicating the streptococcus with benzathine penicillin. In cases of chorea, the treatment varies

Table 1. Update of Jones criteria (2015)[10].

1st RF outbreak:	RF relapse (recurrent RF)
– 2 major criteria; or – 1 major and 2 minor.	– 2 major criteria; or – 1 major and 2 minor; or – 3 minors.
Low risk populations	Risk populations moderate/high
Major Criteria: – Carditis (clinical or subclinical); Arthritis (polyarthritis only); – Chorea; – Erythema Marginate; – Subcutaneous nodule;	Major Criteria: – Carditis (clinical or subclinical); – Arthritis (polyarthritis), polyarthralgia and/or monoarthritis); – Chorea; – Erythema marginatum; – subcutaneous nodule
Minor criteria: – Polyarthralgia – Fever ≥ 38.5°C – Elevation of VHS (≥60mm in the 1ª hour) and/or CRP ≥ 3mg/dL (or greater than the indicated reference value) – Prolonged, age-corrected PR interval (only when carditis is not present).	Minor criteria: – Monoarthralgia – Fever (≥ 38°C) – Elevation of VHS (≥ 60mm in the 1ª hour) and/or CRP ≥ 3mg/dL (or > what reference value is indicated). – Prolonged, age-corrected PR interval (only when carditis is not present).
Proven evidence of previous infection with group-A B-hemolytic Streptococcus (positive oropharyngeal culture, positivity in rapid tests for the detection of streptococcal antigens, high titers of antistreptococcal antibodies).	

in terms of the clinical picture, which may be self-limiting or with moderate to severe symptoms that indicate pharmacological treatment with benzodiazepines, phenobarbital, or antipsychotics. After the treatment of the acute condition, primary prophylaxis is indicated, which corresponds to the administration of antibiotic therapy up to 9 days after the onset of symptoms. Subsequently, secondary prophylaxis should be maintained, both with penicillin as the antibiotic of choice, the duration of which depends on the patient's age, the interval of the last attack, the presence of carditis in the initial attack, the number of relapses, the social condition. and the severity of residual rheumatic heart disease[11]. Both measures aim to reduce disease relapses or progression to cardiac sequelae. In most cases, the CS episode improves within 6 months. A retrospective study was carried out with 90 patients in a tertiary hospital where it was observed that 85% of them had complete remission of motor symptoms within 6 months and another 5% had it after 1 year[12].

CONCLUSION

This report demonstrates the relationship between Sydenham's Korea and late diagnosis of Rheumatic Fever after pharyngotonsillitis caused by group A beta-hemolytic streptococcus, in addition to discussing diagnostic criteria, treatment, prophylaxis and explaining the good prognosis from adequate and early therapeutic management.

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