Sydenham's chorea in Sudan

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Abstract

Background: Sydenham's chorea (SC), is the most common form of acquired chorea in childhood, it is considered as neurological complication of streptococcal pharyngitis. Our aim was to determine the clinical pattern, association of Sydenham's chorea with other manifestation of ARF and the laboratory findings of Sydenham's chorea among Sudanese patients.

Methods: This study is descriptive , retrospective cross- sectional study. Fifty patients with different age groups, were diagnosed as having Sydenham's chorea & followed up at The National center for Neurological sciences, in the period (Jan2017 to Nov2019).

Data were obtained after patients consent of by personal interview or personal review of patients records through a designed questionnaire including demographic data, symptoms, co morbid illness, risk factors, physical examination and related investigations.

Results: The patients' median age was 13.7 years: 88% of the cases occurred between 7-17 years with female predominance(35) . Generalized chorea was seen in 33 (66%) and hemichorea 17 (34%) patients. weakness and hypotonia were common, behavior change (44%), dysarthria (71%), gait change(17.20%)and deterioration of handwriting (13%) Arthritis occurred in (36%), carditis 30 (60%), arthritis and carditis in 18(36%), and pure chorea 14 (28%). Erythema marginatum and subcutaneous nodules were not observed in our patients. Only13 patients (26%) gave a history of pharyngitis.

Conclusion : There were clear evidence of familial predisposition. Sydenham' chorea commonly presents acutely in majority of patients. Chorea firstly appeared with oro-fascial movement followed by limb involvement. Minority of patients gave a history of pharyngitis. Brain imaging was normal.

Background

Sydenham's chorea also known as St. Vitus dance, St. Johannis' chorea, chorea minor, and rheumatic chorea, is one of the major clinical manifestations of acute rheumatic fever (ARF) and it is the most common form of acquired chorea in childhood. It is considered as neurological complication following infection with particular strains of streptococci (i.e., group A beta-hemolytic streptococci^[2] .

The initial illness is usually characterized by a sore throat (pharyngitis) that may be followed, with in approximately 1 to 5 weeks, by the sudden onset of acute rheumatic fever^[4]. The symptom-free period between recovery from pharyngitis to the onset of acute rheumatic fever is known as the "latent period".

In about 20% of patients Sydenham's chorea occurs as the only manifestation of ARF. It is also considered a delayed complication of acute rheumatic fever, occurring as late as 12 months after the initial throat infection. Acute rheumatic fever is an inflammatory disease (i.e., sequelae) following group A streptococcal infection that may affect multiple tissues and organs, including the joints, skin, connective tissues directly beneath the skin (subcutaneous tissues), heart, and brain^[1,2;4] .

Sydenham's chorea may occur, with other symptoms of acute rheumatic fever or as an isolated form. In the isolated form, laboratory evidence of a preceding streptococcal infection may be lacking^[4].

Although more common in children, rheumatic fever can also occur in adults, and migratory arthritis is a common presenting symptom.

These patients need to be considered candidates for prophylactic therapy, as evidenced by an outbreak in 10 young adults in the San Diego Naval Training Camp, three of whom developed valvular damage^[5].

It is a movement disorder characterized by chorea, emotional lability, and hypotonia. Emotional changes manifest themselves in outbursts of inappropriate behavior, including crying and restlessness. In rare cases, the psychologic manifestations may be severe and may result in transient psychosis^[6].

Methods

Study design:

This study is descriptive and retrospective cross-sectional study.

Study Time:

This study was conducted between January 2017 and November 2019

Study area:

The national centre for neurological sciences Khartoum state; a tertiary hospital for the referral of all neurological cases in Sudan

Study population:

Sudanese patients who were diagnosed as having Sydenham's chorea at Alshaab teaching hospital – neurology department in the period (Jan 2017 to Nov. – 2019)

Diagnostic Criteria:

The diagnosis of Sydenham's chorea has been Establishing aided by judicious application of the Jones criteria; included three components:

major manifestations

- Carditis
- Polyarthritis
- Chorea

- Erythema marginatum
- Subcutaneous nodules

1. minor manifestation;

- Arthralgia
- Fever
- Elevated acute phase reactants (erythrocyte sedimentation rate, C-reactive protein)
- Prolonged PR interval

2. Evidence of a preceding group A streptococcal infection

- Elevated or rising streptococcal antibody titer, most often Antistreptolysin O titre.

Study size & sampling technique:

Fifty patients with different age groups, representing the whole patients who were diagnosed as having Sydenham's chorea & followed up at Alshaab teaching hospital, neurology department .

Inclusion criteria:

- Sudanese patients (adults & children)
- Males & females
- Symptoms & signs of Sydenham's chorea.

Exclusion criteria:

- Non Sudanese patients
- Patients with other voluntary movement disorders or undetermined type of movement disorders.

Data collection:

Data were obtained after patients consent of by personal interview or personal review of patients records.

Study tools:

A retrospective collection of data from the patients with Sydenham's chorea or their records through a designed questionnaire including demographic data, symptoms, co morbid illness, risk factors, physical examination & related investigations.

Echocardiography and electrocardiography were performed in all patients, brain image was done in 11 patients.

Analysis:

All collected data were entered into the computer using the statistical package program for social science (SPSS) to analyze the data via simple descriptive statistics. (Analyzer is specialized personnel in SPSS).

Results

This study is descriptive and retrospective cross-sectional study in Sudanese patients with Sydenham's chorea.

Sydenham's chorea can present at different age group, the majority of the cases occurred between 7 -17 years. The patients' median age was 13.7 years. In this study, 50 patients were diagnosed to have Sydenham's chorea, among them 35 patients (70%) were females and 15 patients (30%) were males. There was positive family history of acute rheumatic fever in 34% of the studied group. The chorea lasted from 2 to 4 weeks in 26 patients (52%), 5 to 10 weeks in 19 patients (38%) and 11 to 15 weeks in 5 patients (10%).

In all patients the movements were suppressed during sleep (100%) and this was being the provocation factor 12 patients (25%).

Sydenham's chorea have acute onset in 34 patients (68%) ,while 16 patients (32%) had gradual onset .

Generalized chorea was seen in 33 patients(66%) and hemichorea 17 patients (34%) . It was seen that 41 patients (82%) had their chorea initially started with limb involvement , while 9 patients (18%) had their chorea firstly appeared with orofascial movement followed by limb involvement and weakness occurred in 19 patients (38%), hypotonia in 23 patients (46), dysarthria 35 patients (70%), gait change 9 patients (18%) and deterioration of handwriting 6 patients (12%). Darting tongue and milking sign were found in 21 patients (42%) ,emotional liability was found in 26 patients (52%) followed by restlessness 22 patients (43%) while depression occurred in patients 2 patients (5%).

The association of Sydenham's chorea and other manifestations of acute rheumatic fever :

The association of Sydenham's chorea and other manifestations of acute rheumatic fever, carditis occurred in 30 patients (60%), 18 patients had arthritis only (36%), arthritis and carditis in 18 patients (36%), and pure chorea 14 (28%). Erythema marginatum and subcutaneous nodules were not observed in our patients.

Regarding investigations, Elevated ASO levels were detected in 44 patients (88%). Only 13 patients (26%) gave a history of pharyngitis. Highly elevated ESR was detected (>100 mm/h) in 18 patients (36%) and moderately elevated ESR (30-70 mm/h) in 32 patients (64%) patients. Prolonged PR (or first-degree block) was noted in 9 patients (18%) . Mitral regurgitation was found in 20 patients (40%) and combination of mitral and aortic regurgitation occurred in 10 patients (20%). Brain imaging, which was performed in only 11 patients, was evaluated as normal in all.

Ten of 50 patients (20%) had a recurrent attack; all of them were the second attack. The interval between attacks varied from 2-3 years. Carditis was detected in all patients with recurrences

Discussion

This study revealed that 35 (70%) of the patients were female and 15 (40 %) patients were male. Females were found to be most frequently affected than males. The mean age incidence of patients of Sydenham's chorea was found to be 13.7, the median age range between 7 to 27 years.

In Australia study the female percentage was (89.8 %) & the mean age was 10.9 years at first episode [7]. In the Indian study girls incidence was (60 %); the mean age at presentation was 11.1 yr (range 7-16 yr) [8].

The Turkey study revealed that, the mean age at the onset of the symptoms was 11.7 +/- 2.6 years (range 6-17 years). The study revealed that 63% of the patients were female and 37% were male (male/female: 1.7/1) [9].

In comparison to this study confirmed that the percentage of our female affection predominated the male affection.

The mean age in our study is higher than that found in other studies; this may reflect that, Sydenham's chorea occurs at higher age group in Sudanese in comparison to other population or it can be attributed to second attack of Sydenham's chorea considering that Sydenham's chorea occasionally presents a subtle form.

Much of the studies documented that Sydenham's chorea is still a common problem in the developing countries due to the high incidence of ARF; the Assiut, Egypt study found that rheumatic chorea was most common among the studied population [10].

The chorea last from 2 to 15 weeks in the study, this finding is in line with the observations of others [7]. Regarding the pattern of onset of chorea most patients were found to have acute onset (67.30%) rather than gradual (32.70%), this is supported by the fact that Sydenham's chorea typically has usually acute onset.

Generalized chorea in patients with Sydenham's chorea was seen in 33 (66%), hemichorea in 17 patients (34%), comparing this with India study, generalized chorea was seen in 40 (66.6%) and hemichorea in 20 patients (33.3). This reflects that Sydenham's chorea commonly present as generalized rather than partial chorea.

The study revealed that most movements initially started at the limbs (82%) rather than orofacial (18%). This is described well in the literature.

The study found that chorea suppressed during sleep (100%) & provoked with certain factors e.g. stress (25%) & sudden movement (7.7%). This matches most of the available reports^[11,12].

Higher function disturbances were common; of which speech was the most affected item, mainly as dysarthria (71.1%); this finding correlates with the review, that chorea commonly associated with dysarthria.

Most of the patients who diagnosed as Sydenham's chorea exhibited decreased school performance (27 patients – 53.8%), emotional lability (34 patients – 69.2%) this goes with what has been found in literature^[6,13,14]. Darting tongue & milking sign were found in 42.3% of all patients.

Psychiatric symptoms were reported in 23 patients (44.2%) mainly as restlessness & emotional lability. This confirmed well with the literature^[5] which reported high frequency of these symptoms in Sydenham's chorea.

In our study, past history of ARF occurred in 12 (24%) patients, in comparison to the Turkey study a past history of ARF^[9] was accounted for 30%. Family history of ARF occurred in 34% in this study; this is similar to the results of other studies, such as Sydenham's chorea study in western Pennsylvania in which family history of ARF existed in 30% of patients^[11].

The study found ten patients (20%) had a recurrent attack; the recurrence rate was 37.9% in the Turkey study; this can be explained by the short time of our study & the lack of close follow up to study sample for long period.

The duration of Sydenham's chorea varied between 7 to 50 days in our study; closely related to other reports. All the patients with Sydenham's chorea had their movement stopped completely within 1 to 2 weeks after treatment, no single case persisted, this goes with the literature as persistence rate in patients with is low in Sydenham's chorea^[3,5].

To discuss the association of Sydenham's chorea with the other manifestations of ARF, the results are as following; Eighteen patients had arthritis only (36%), carditis 30 patients (60%), arthritis and carditis in 18 patients (36%), and pure chorea 14 patients (28%).

Erythema marginatum and subcutaneous nodules were not observed in our patients these are comparable to the Turkey study; in which Carditis was 25%, arthritis in 33%, carditis & arthritis in 28%, carditis & chorea in 12%, and chorea in 3%; also we can see carditis (44%), arthritis (11%), erythema marginatum (3%), and subcutaneous nodules (0%) were collected from Sydenham's chorea study in western Pennsylvania^[11].

The study showed carditis occur in 30 patients (60%). six patients (12%) with pure chorea had silent valvitis demonstrated only by echocardiographic testing but without any significant murmur (mitral

regurgitation in 1 and combination of mitral and aortic regurgitation in 3 cases). So carditis increased up to 72%, reflects high affection (more severe disease) in Sudanese patient.

In our study Only 13 patients (26%) gave a history of pharyngitis ,this is because pharyngitis may precede the onset of chorea by as much as 6 months, many patients may not provide a history of streptococcal infection, but still we have allow percentage as history of pharyngitis is obtained in 41%, in Turkey study.

Elevated ASO levels were detected in 44 (88%) of the patients. Highly elevated ESR was detected (>100 mm/h) in 18 (14%) and moderately elevated ESR (30-70 mm/h) in 77 (59%) patients. This reflected that acute phase reactants were raised almost in most of the patients with Sydenham's chorea. In comparison to other studies in Turkey Elevated ASO titre in 83%; in Indian study; a raised ESR and positive ASO titer were seen in 37 and 20 patients respectively.

Prolonged PR or first-degree block was noted in 9 (18%) patients in our study & was found to be prolonged in 13% of patients in India one.

Mitral regurgitation was found in 20 patients (83%), aortic regurgitation in 6 patients (44%) and combination of mitral and aortic regurgitation in 10 patients (27%). The mitral valve was the most commonly affected valve (64%), followed by aortic valve (22%). The mitral valve is the most commonly affected valve in rheumatic heart disease.

In 50 patients with Sydenham's chorea, brain image(brain MRI) was done in 11 patients, all was reported to be normal, this goes with most of the studies done for that issue, one of these was done in western Pennsylvania which revealed that in patients with Sydenham's chorea, brain computed tomography was abnormal in just 1 of 20 patients and that the abnormalities did not aid in diagnosis and included nonspecific increased finding ^[11].

Conclusion

The mean age of presentation of Sydenham's chorea was between 7-17 years. The study showed predominant female preponderance. The study showed clear evidence of familial predisposition. Sydenham's chorea commonly presents acutely in majority of patients Invariably in all cases the movements were suppressed during sleep, stress being the most provocational factor.

Chorea firstly appeared with orofascial movement followed by limb involvement. Darting tongue & milking sign were common. Generalized chorea was more prevalent than hemichorea Psychiatric symptoms manifested as emotional liability restlessness and depression. Eighteen patients had arthritis only (36%), carditis 30 (60%), arthritis and carditis in 18(36%), a pure chorea 14 (28%). Erythema marginatum and subcutaneous nodules were not observed in our patients. Elevated ASO levels and ESR were detected in most of the patients, minority of patients gave a history of pharyngitis. Prolonged PR or first-degree block was noted.

The mitral valve was the most commonly affected valve followed by aortic valve. Brain imaging, which was performed in a minority of cases, was evaluated as normal.

Recommendations

As Sydenham's chorea is a common disease in Sudan, further studies are needed to be conducted in this direction. All patients with chorea should be screened for Sydenham's chorea. Long-term adherence to secondary prophylaxis is crucial following all episodes of acute rheumatic fever, including chorea, to prevent recurrence.

Declarations

Availability of data and materials

The materials datasets used and/or analyzed during this study are available from the corresponding author on reasonable request.

Ethical Considerations

Ethical approval was obtained from State Ministry of Health, ethical committee.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

All authors participated in planning the study, data collection, results and discussion sections.

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