Brain & Development 38 (2016) 827–834

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Original article

Predictors of recurrence in Sydenham's chorea: Clinical observation from a single center

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Received 8 February 2016; received in revised form 14 April 2016; accepted 19 April 2016

Abstract

Objective: Sydenham's chorea is the most common cause of acquired chorea in children and is the major manifestation for acute rheumatic fever. Despite being known as a benign, self-limiting condition, recurrences and persistence of symptoms can be seen. In this study, we aimed to evaluate retrospectively the clinical and laboratory features of patients with Sydenham's chorea and the rate and the course of recurrences, and to assess the risk of recurrences.

Methods: The study was a retrospective study conducted in a tertiary hospital. Patients with Sydenham's chorea who were admitted to our outpatient clinics between January 2013 and June 2015 were included. Both newly diagnosed and follow-up patients were enrolled during this period. We retrospectively reviewed the medical charts of the patients.

Results: There were 90 patients with female predominance. The mean age of onset was 11 ± 2.4 years. Complete remission was maintained in 77 patients (85.6%) at 1–6 months and 4 patients had symptoms at more than 12 months. Patients were followed for 6 months to 9 years. The recurrence rate was 16%. When we compared recurrent patients with the non-recurrent group, complete remission in 6 months, the presence of persistent chorea, and regular use of prophylaxis were significantly different between the 2 groups.

Conclusions: Sydenham's chorea is still an important health problem and has high morbidity in patients with recurrent and persistent chorea. The irregular usage of antibiotic prophylaxis, failure to achieve remission within 6 months, and prolongation of symptoms for more than 1 year are risk factors for recurrence of chorea.

Keywords: Sydenham's chorea; Rheumatic fever; Recurrence

1. Introduction

Chorea is a movement disorder characterized by rapid, involuntary, irregular, jerky movements affecting the limbs, face, and trunk. There are many causes of childhood chorea but Sydenham's chorea (SC) is the most common cause of acquired chorea in children [1–3]. SC is a major manifestation for acute rheumatic fever (ARF), caused by group A β-hemolytic *Streptococcus* infection [4].

The incidence and prevalence of ARF have been decreasing in developed nations since the early 20th century due to improved living conditions and the use of antibiotics for streptococcal pharyngitis, but ARF

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http://dx.doi.org/10.1016/j.braindev.2016.04.010

continues to be a major cause of morbidity and mortality among young people in developing countries [5].

SC is caused by antibodies against group A β-hemolytic *Streptococcus* bacteria. The antibodies, which are called anti-basal ganglia antibodies, cross-react with the neurons within the basal ganglia [6]. Cerebral arteritis with cellular degeneration develops. Endothelial swelling, perivascular round cell infiltration, arteritis, and petechial hemorrhage are found on histological examination [7,8]. These changes result in basal ganglia dysfunction. Involvement of the putamen causes motor signs and behavioral changes occur via caudate involvement and cortical dysfunction [2].

SC is characterized by choreiform movements, hypotonia, muscle weakness and neuropsychiatric signs, including obsessive–compulsive signs, emotional lability, anxiety, attention deficit, tic disorders, executive function disturbances and psychotic features [2,9]. The disease usually has a monophasic course but recurrences can be seen in some patients several months after the initial attack [10].

In this study, we aimed to evaluate retrospectively the clinical and laboratory features of patients with SC who were admitted to our clinic. We also evaluated the rate and the course of recurrences and assessed the risk of recurrence.

2. Materials and methods

The study was a retrospective study conducted in a tertiary hospital in Ankara, Turkey. Patients diagnosed with SC who were admitted to our pediatric outpatient neurology clinics between January 2013 and June 2015 were included. Both newly diagnosed and follow-up patients were enrolled during this period. We retrospectively reviewed the medical charts of the patients. The diagnosis of ARF was based on the updated Jones criteria [4].

In our pediatric outpatient neurology clinic, there is no standardized protocol for evaluation of patients with chorea; a detailed history and physical and neurological examinations were performed on all patients. All patients underwent routine blood tests including total blood cell count, serum electrolytes, glucose level, renal and hepatic function, anti-Streptolysin O (ASO) titers, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), ceruloplasmin, thyroid function tests, antinuclear antibodies, antiphospholipid and anticardiolipin antibodies, rheumatoid factor, and throat culture. Cardiac evaluation was done by pediatric cardiologists and all patients underwent electrocardiography and echocardiography. Brain magnetic resonance imaging (MRI) was performed if necessary. All data were saved as a structured file by the pediatric neurologist at each visit. We retrospectively reviewed these data, which included sex, current age, age at chorea presentation, duration of chorea until admission, duration of improvement after the first attack, duration of follow-up period, bodily distribution of chorea, presence or absence of carditis as detected by echocardiography, recurrences, usage of prophylactic antibiotics after diagnosis of SC, antichoreic treatment, and brain MRI findings (if performed).

Evaluation of the patients was not standardized because patients were evaluated by different pediatric neurologists. Patients were generally evaluated once a week until chorea was controlled. After control of chorea was achieved, the patients were followed monthly/bimonthly. During reduction of symptomatic medication, patients were evaluated every 1–2 weeks. After the end of treatment, patients were followed up at 3-month intervals for the first year and then twice a year.

We classified the chorea according to localization (hemichorea, movements affecting one side of the body; generalized, movements affecting the whole body) and severity of the symptoms (mild, moderate, and severe). Symptoms were classed as mild in the presence of minimal movements, moderate in the presence of movements of obvious discomfort to the patient, and severe if there were movements sufficiently incapacitating for the patient to require assistance with daily activities [7]. We could not grade the degree of chorea according to the Universidade Federal de Minas Gerais Sydenham's Chorea Rating Scale because of the retrospective nature of the study and lack of data [11]. We also grouped the patients according to transitory or persistent chorea. Persistent chorea was defined as symptoms occurring for more than 1 year [12].

We recorded the ASO titer, ESR, and CRP level of each patient from samples obtained during the first attack when the patients had recurrent or persistent abnormal movements. An ASO titer >200 IU/ml, ESR >20 mm/h, CRP >0.8 mg/dl were defined as increased.

Recurrence was defined as the development of new signs lasting more than 24 h and separated by a minimum of 2 months from the previous episode [10]. At the time of recurrence, patients were assessed for rheumatic fever activity.

Statistical analysis was performed on a personal computer using SPSS/PC version 15.0. Differences in proportions were assessed by χ^2 test or Fisher exact test, as appropriate. The Mann–Whitney U test and Student's t test were used for analysis of data between the groups. $P < 0.05$ was considered statistically significant.

3. Results

A total of 90 patients were enrolled in the study; 68.9% ($n = 62$) were female. The mean age of chorea onset was 11 ± 2.4 years (range, 4.5–16 years). Males were slightly younger (10.5 ± 2.2 years) at chorea onset