International Journal Of

Recent Scientific Research

ISSN: 0976-3031
Volume: 7(4) April -2016

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THE OFFICIAL PUBLICATION OF
INTERNATIONAL JOURNAL OF RECENT SCIENTIFIC RESEARCH (IJRSR)
http://www.recentscientific.com/ recentscientific@gmail.com

ISSN: 0976-3031

Available Online at http://www.recentscientific.com

International Journal of Recent Scientific Research
Vol. 7, Issue, 4, pp. 10348-10350, April, 2016

International Journal of Recent Scientific Research

Case Report

ISOLATED CHOREA A RARE MANIFESTATION OF RHEUMATIC FEVER

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ARTICLE INFO

Article History:

Received 20th January, 2016
Received in revised form
29th February, 2016
Accepted 30th March, 2016
Published online 28th April, 2016

ABSTRACT

Acute rheumatic fever is an autoimmune disease that occurs approximately 3 weeks after an untreated group A beta hemolytic streptococcal sore throat infection. Sydenham's chorea is an enigmatic and may appear as an isolated manifestation or precede or accompany an acute attack associated with other criteria of the disease. Here we describe two cases of isolated chorea.

INTRODUCTION

The disease was first named by Thomas Sydenham in 1686 as 'St Vitus Dance' to differentiate it from dancing mania. It is a rare manifestation, affecting less than 5% of patients and occurs among patients between the age 5 and 15 years, Table 1 predominantly in females and usually 1-6 months after the onset of sore throat [1]. It is an autoimmune disease following infection with group A beta hemolytic Streptococci as its M proteins induce production of antineuronal antibodies those cross react with the body owns cells in the basal gangli, resulting into involuntary movements and psychiatric disturbances[2]. Sydenham's chorea is characterized by distinctive symptom complex of involuntary, purposeless, rapid movements that are often associated with incoordination, muscle weakness and behavioral abnormalities [3]. It is a major manifestation of rheumatic fever and, according to the latest modification of the Jones criteria in 1992, a criterion sufficient even by itself for the diagnosis of disease [4].

Table 1 Clinical classification of Sydenham chorea [2]

Mild	Presence of minimal movements of the body.
Moderate	Movements of the body of the patient are inconveniencing but do not interfere with daily activity.
Severe	Abnormal movements of the body sufficiently enough for the patient to require assistance for running daily activity

With widespread availability and use of antibiotic for streptococcal A infection under ARI control Programme in our country, Sydenham chorea has become rare.

Case 1

A 9 years old male was admitted at a largest teaching tertiary care centre of central India with history of sore throat 3 months before and was treated by private practioner. Since last 2 months, he had purposeless, rapid, uncontrollable writhing movements of limbs, head, neck and face and such movements were aggravated by activity and disappeared in sleep. Initially school teacher had noticed that he was poorly concentrating in studies and was unable to hold a pen and had difficulty in writing. Mother also observed that he had poor speech with good comprehension but fluency was poor. He had no past history of any cardiac complaints, arthritis and rash or nodule. Examination revealed patient with rapid, uncontrollable jerky movements of limbs, face, neck, head and paucity of speech with no abnormality on systemic examination. The two dimensional echo finding showed normal myocardial wall motion with mildly dilated left atrium, mildly thickened posterior valve leaflet without pericardial effusion or vegetation. A Doppler study shows mild mitral regurgitation. Chest radiograph was within normal limit. Electrocardiography shows nonspecific ST/T changes. CT scan head and Electroencephalogram yielded no abnormality. Complete blood count, blood sugar, serum electrolytes, serum ceruloplasmin, urine analysis were within normal limit.

Based on echo finding, abnormal movements and history of sore throat, the diagnosis of Sydenham's chorea was made. Patient was treated with haloperidol, antibiotics and salicyclates. Symptoms resolved within two weeks of admission. He was discharged and put on Benzathine penicillin prophylaxis and he followed up for next six months. There was no recurrence of chorea.

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Case 2

A 11years old female adolescence presented with purposeless, jerky, uncontrollable involuntary movement of left upper and lower limb with slowing of speech and verbal response. The involuntary movements were severe enough to impair her daily activities such as writing, brushing teeth and holding food bowl. She had an episode of fever, mild arthralgia and sore throat one month before. Physical examination reveals that she was afebrile, oriented and vitals were stable. Involuntary movement was present as describe above. The muscle tone, deep tendon reflex and muscle power were all symmetrical and normal. Cardiovascular examination showed normal heart sound with no murmur. Blood test shows normal electrolytes, calcium and complete blood count. ESR was 50mm at end of first hour. ECG shows prolonged PR interval CT scan head, Nerve conduction study and EMG were within normal limit. The clinical and laboratory finding were compatible with diagnosis of Sydenham's chorea. Patient was treated with Penicillin, Aspirin and Haloperidol. Clinical improvement was observed and she was discharged on 15th day of treatment. Patients was put on Benzathine penicillin prophylaxis and asked for follow up. During subsequent follow up, she had no recurrence of chorea.

DISCUSSION

The incidence of rheumatic fever has declined in the developed countries may be because of improvement in living standard, overall socioeconomic development and the wide availability of penicillin. Nevertheless, it is still a major health problem in developing countries, where it and its sequelae account 25-40% of all cardiac admissions. [1]. Sibling studies have demonstrated an inherited susceptibility to certain patterns of rheumatic fever. Taneja *et al* and Kaur *et al* had identified B lymphocyte surface" rheumatic" antigens among patients of rheumatic fever. These findings suggested that there could be genetic susceptibility to rheumatic fever [5, 6]. Arthritis is the most (80%) manifestation of acute rheumatic fever followed by carditis (40-75%), erythema marginatum and subcutaneous nodules [1]. Sydenham's chorea though it is rare affecting less than 5% of patient but it is a major manifestation and with the 1992 modification of the Jone's criteria , is sufficient alone to make the diagnosis of rheumatic fever. A 35-65% cases isolated chorea was reported from Brazil and Malaya [3, 7]. Unlike acute rheumatic fever, the incidence trend of isolated chorea fluctuated with peaks every few years.

Sydenham's chorea was first named by Thomas Sydenham in 1686 as 'St. Vitus Dance' to differentiate it from dancing mania. The association between Sydenham's chorea and arthritis was established by Stroll in 1780. Our one case had also arthritis with chorea. Various studies reported, the mean age of chorea is ranging from 10-11.5 years and youngest one is 7 years old female reported by Kulkarni from India [4]. Our cases are also falling into same age group. Female predominance is noted in almost all studies of chorea and this sex disproportion was evident in adolescence and it suggested that sex hormone play a role the sex difference. The main feature of Sydenham's chorea is involuntary movements. These can be generalized or unilateral. These movements occur at rest, may start suddenly or gradually, and are exacerbated by stress. They disappear during sleep. Usually the patient has abnormal neurological signs with hypotonia and motor restlessness which can lead to coordination problems, gait disturbances and speech impairment. As a result, the activities of daily living can be severely disrupted. Such patients can have psychological and psychiatric manifestation such as emotional lability, anxiety, personality changes and depression also. Our cases had also similar presentation. Since chorea is generally a late manifestation, it is unusual to find clinical or even immunological (laboratory) evidence of streptococcal infection, and the erythrocyte sedimentation rate is usually normal. Recurrent attacks of chorea are not uncommon but recurrence many years after the initial attack are rare and suggest late chorea may be due to reactivation by other mechanism. Drugs like phenytoin, oral contraceptive and pregnancy may precipitate late chorea [4].

Sydenham's chorea, once considered as a self limiting, is now felt to require more aggressive treatment because it can cause great functional impairment to patient. There are several pharmacological agents included in treatment like neuroleptics (haloperidol, trifluoperazine, chlorpromazine), levodopa, hydroxyzine, phenobarbital, carbamazepine. Our cases are well responded to haloperidol without observable side effect. Haloperidol acts by blocking the dopamine receptor. Recently, valproic acid has been advocated to be effective in treating chorea cases. Immunological therapy with steroids or intravenous immunoglobulin had been tried to treat Sydenham's chorea but the response was not satisfactory. Further controlled trials are needed to investigate their efficacy [1,4]. We treated our cases with penicillin for eradicating streptococci,

Aspirin as a anti-inflammatory agent in mild carditis and arthrtis and haloperidol for involuntary movement. Our patient responded well without observable side effect. The American Heart Association recommended prophylaxis to be continued at least 10 years after the last episode of rheumatic fever or until the patient are well into adulthood [8]. We also recommended Benzathine penicillin prophylaxis to our case and ask them to continue for recommended period.

CONCLUSION

Sydenham' chorea is a rare presentation of acute rheumatic fever. Treatment with haloperidol may be useful for those having difficulty with their activities of daily living. Long term follow-up and antibiotic prophylaxis are required to prevent recurrence of rheumatic fever.

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How to cite this article:

Rajkumar M. Meshram., Abhisheik S and Hina Agrawal.2016, Isolated Chorea A Rare Manifestation of Rheumatic Fever. *Int J Recent Sci Res.* 7(4), pp. 10348-10350.

TSSN 0976-3031