Sydenham's Chorea with Silent Cardiac Lesions, Mimicking Encephalitis in a 13 Year Old Girl

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ABSTRACT

Sydenham's chorea is an uncommon neurological manifestation of rheumatic fever and has many and varied differential diagnosis. It may mimic encephalitis when presents as an isolated feature even when silent cardiac lesions are present. Early diagnosis, treatment and penicillin prophylaxis prevents recurrence and progression of cardiac lesions. Prompt symptomatic relief and alleviation of distress is obtained with therapeutic intervention. A case of rheumatic chorea with silent cardiac valve lesions which mimicked herpes simplex encephalitis with choreoathetosis, in a 13 year old girl is presented along with review of literature.

KEY WORDS

Chorea, Encephalitis, Rheumatic fever, Silent carditis

INTRODUCTION

Sydenham's chorea is a neurological manifestation which occurs in 20-40% of acute rheumatic fever.[1] It may present as an isolated feature or along with other manifestations of rheumatic fever.[2] It is a delayed manifestation and other symptoms may subside when it appears, usually up to 6 months after Group A β-hemolytic streptococcal infection.[3] Isolated attacks without other rheumatic fever symptoms, are common in recurrent chorea.[1] However, concomitant carditis may occur in up to 80%.[1] As chorea is an uncommon presentation, it has many and varied differential diagnosis and the cardiac lesion may be overlooked. Sydenham's chorea is important to diagnose because early treatment and penicillin prophylaxis minimize recurrence and cardiac damage. We present here a case of Sydenham's chorea in a 13 yr old girl with silent cardiac valve lesions, whose presenting features mimicked herpes simplex encephalitis.

CASE REPORT

Cardiac findings were normal and unremarkable. Laboratory investigations showed: Total leukocyte count 18 x 109/L, Erythrocyte sedimentation rate 47 mm / hr, C reactive protein 110.77 mg/L, normal fasting blood glucose and thyroid-function tests. Serology was negative for herpes simplex virus. The antistreptolysin O titre was 400 IU/mL (reference value < 200 IU/mL), indicating a recent streptococcal infection. Throat culture revealed growth of Streptococcus pyogenes. ECG showed prolonged PR interval and echocardiography with Doppler studies reported various grades of mitral, tricuspid and aortic regurgitations. Ophthalmic evaluation showed no Kayser-Fleischer rings, On the basis of these clinical presentation and laboratory findings, our patient was diagnosed as Sydenham's chorea with carditis.

Treatment was started with oral amoxicillin for 10 days, followed by prophylactic benzathine G penicillin. She responded steadily to oral aspirin with gastric prophylaxis. For symptomatic control of choreiform movements haloperidol was given in low doses. She improved progressively and was discharged on day 14. On follow-up visits at monthly intervals, no recurrence of symptoms and progression of cardiac lesions were noted after two months. She appeared active and cheerful with normal speech and gait.

A 13 year old girl from a distant rural district was brought to emergency department of KMCTH with 10 days' history of sudden onset of irritability and involuntary movements of upper limbs. She had irrelevant vocalizations and was confused and unstable to walk. She had emotional lability and was unable to feed by herself. She had sore throat two weeks earlier which subsided without treatment. On examination, she was febrile and confused with equivocal signs of meningeal irritation. She was admitted to pediatric ward with a provisional diagnosis of herpes simplex encephalitis with choreoathetosis.

The involuntary movements disappeared during sleep. Her cranial CT scan and CSF examination reports were normal. Meanwhile, her emotional lability became more marked. Further examination revealed generalized weakness, congested pharynx and hypotonia. Hand pronator sign (Fig. 1), hand grip milkmaid sign (Fig. 2) and darting tongue sign were positive.

Figure 1. Showing positive hand pronator sign

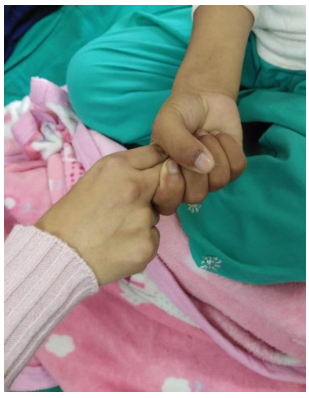

Figure 2. Showing positive hand grip milkmaid sign

DISCUSSION

Our patient at presentation, had fever with irritability and involuntary movements. Later, emotional lability, confusion and irrelevant vocalizations were noted, mimicking encephalitis. In the literature, involuntary movement (choreoathetosis) has been reported after herpes simplex encephalitis with basal ganglia involvement.[4] CSF analysis and neuroimaging led to diagnosis. In our patient CSF findings and CT scan report were normal.

A similar presentation as in our patient, with acute onset of involuntary movements in a nine year old girl with silent rheumatic cardiac valvulitis has been reported from Hongkong.[5]

The latent period following an episode of sore throat in rheumatic chorea, is usually longer. Shorter duration apparently led to diagnostic pitfall in our patient. But, additional features like motor hyperactivity, irritability and increased emotional lability present in our patient were also found in all the patients in a series of eleven cases reported by Swedo et al.[6] Obsessive-compulsive symptoms predominated in their patients.

In our patient, clinically silent cardiac lesions formed a major criteria for rheumatic fever. Such concurrent cardiac lesions have been documented in up to 80% of isolated rheumatic chorea patients by echocardiography with Doppler, as recommended in all confirmed and suspected cases of acute rheumatic fever.[7]

Incidence of chorea, predominates in pubertal girls, which conforms with our patient. Chorea is a major manifestation of rheumatic fever in the revised Jones criteria, 2015.[8] It was first described by Thomas Sydenham in 1686 and was found as an isolated feature in 12% cases of acute rheumatic fever in a case series of 134 patients.[9]

Rheumatic fever cases have been reported as diagnosed after features of chorea became evident. However, cardiac involvements were found in isolated chorea during recurrence.

In chorea, lesion is present in basal ganglia, resulting from the immune responses against group A streptococcal infection by the host. Anti-neuronal antibodies were found in the basal ganglia of the brain in such cases. Mimicry was demonstrated by monoclonal antibodies (mAbs) from rheumatic chorea patients between lysoganglioside (present in rheumatic chorea) and the group A streptococcal epitope (N-acetyl-glucosamine).[10]

The treatment course in our patient was smooth. Oral haloperidol was well tolerated. No adverse effect was noted throughout hospital stay and during follow-up. Although, regarded as a drug of choice for the symptomatic control, haloperidol has the potential for significant side effects needing titration and dose adjustment.

Besides, corticosteroids, valproate and immunoglobulin have also been described as other options Rheumatic chorea has good prognosis with complete remission of motor symptoms reported in 85% by 6 months, and additional 5% by end of one year in a retrospective study of 90 patients.[11] Our patient had no recurrence at follow-up two months after discharge.

Isolated rheumatic chorea may mimic herpes simplex encephalitis with choreoathetosis causing diagnostic indecision. Concurrent silent cardiac valve lesions may be present. Penicillin prophylaxis is essential to prevent recurrence of chorea and progression of cardiac lesions.

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