CASE REPORT

Chorea gravidarum: a rarity in West still haunts pregnant women in the East

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SUMMARY

A pregnant woman, in her early 20s, presents with chorea following an emotional outburst. While the family members feel it to be a spirit haunting her, we try to establish the medical diagnosis of the present condition. There is a history of rheumatic fever in the past and examination reveals choreoathetoid jerky movements of her upper limbs with jerky speech, spooning of her limbs and demonstration of milkmaid's grip. Laboratory investigations did not reveal anything interesting. We discuss the diagnosis and management of this patient.

BACKGROUND

Chorea gravidarum is a rare movement disorder of pregnancy. With the declining incidence of rheumatic fever, chorea underlying rheumatic heart disease has become a rarity in the developed world. However, isolated cases still occur in the developing countries and obstetricians need to be aware of this rare manifestation. Though the exact aetiology remains unknown, prompt diagnosis and treatment reduces maternal morbidity and prevents any complication. This is a case of chorea gravidarum in third trimester of pregnancy with a normal fetal outcome.

CASE PRESENTATION

A 21-year-old primigravida of low socioeconomic status from distant hilly area of Garhwal presented to the outpatient department at 36 weeks' gestation with violent abnormal movements of her upper limbs for 4 days. The history was elicited from her mother-in law who told that patient started having abnormal movements of her tongue which progressed to involve her both arms in the time span of a day when she heard the news of her father's death. The involuntary movements in her tongue and arms were neglected as the attendants took her to a local priest thinking it as an evil spirit in the hills. Her movements disappeared when she slept for initial 2 days but they had become more violent since last 2 days so much so that she could not sleep. The patient herself complained of excessive salivation and she denied any movement disturbance. She had no antenatal health check-ups in this pregnancy. The first and second trimesters of her pregnancy had been uneventful. Three weeks prior to admission in our hospital, she had fever, sore throat and migrating polyarthralgia involving her left wrist followed by left knee joint. Her symptoms had subsided after taking some antibiotics and analgesics from a local health centre. She reported history of similar episode of fever associated with

joint pain in the left wrist and right knee, which was relieved spontaneously in 2 days and was followed by pain in left elbow joint (fleeting joint pain) at the age of 15 years. This episode was preceded by sore throat for around 10 days. This history was suggestive of an episode of rheumatic fever. However, she denied any history of taking long-term injectable penicillin for the same. There was no history of psychiatric illnesses in the past. There was also no history of any neuroleptic drug or metocloperamide intake.

On examination, patient was right-handed, afebrile, pale, conscious, oriented, emotionally labile but depressed. The tongue was darting even at rest without patient being able to control it. She had choreoathetoid, purposeless, irregular and jerky movements bilaterally in her upper limbs. The patient was constantly trying to cover her head with her cloth each time she was having choric movements. The patient was fidgety. Her speech was 'jerky' with sudden changes in pitch and loudness. On holding the arms outstretched, there was hyperextension of the fingers with dorsiflexion of the wrist, also known as 'spooning' (figure 1). On asking the patient to squeeze the examiner's finger, the pressure of the patient's grip was increasing and decreasing continuously and capriciously, confirming the presence of 'milkmaid's grip' or 'milking sign'. Tone was slightly reduced in upper limbs than lower limbs. Power and reflexes were normal in all four limbs. Sensory system and all cranial nerve examination were normal with flexor plantar response. There were no cerebellar signs and signs of meningeal irritation. There was no family history of any neurological disease. Abdominal

Figure 1 Demonstration of 'spooning' in outstretched hands.

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examination revealed uterine size corresponding to 34 weeks gestation, with cephalic presentation.

INVESTIGATIONS

Ultrasound of the abdomen revealed gestational age of 32 weeks 4 days with cephalic presentation. Laboratory studies were as shown in table 1.

Imaging studies, including two-dimensional ECG and CT brain were found to be normal. Slit-lamp examination of the eyes was normal.

DIFFERENTIAL DIAGNOSIS

The differentials which were considered on history and examination were chorea gravidarum, systemic lupus erythematosus, primary anti-phospholipid antibodies, viral encephalitis, Huntington disease and Wilson disease. The negative reports of laboratory investigations of the antinuclear antibodies and C reactive protein helped in excluding systemic lupus erythematosus and primary antiphospholipid antibody. The absence of associated symptoms of fever, coryza helped in excluding viral encephalitis. There was no family history of Huntington disease and disease progression was also not suggestive of Huntington disease. The slit-lamp examination and copper estimation helped exclude Wilson disease. The positive history of rheumatic fever in the past and precipitation of these symptoms during pregnancy indicated towards the likelihood of chorea gravidarum which was rheumatic in origin.

TREATMENT

The patient was admitted in a quiet isolated ward and given one dose of benzathine penicillin, iron and calcium supplements were given along with multivitamins. After proper hydration and rest, her involuntary movements were markedly reduced over a period of 4 days stay in the hospital. Attendants were advised to keep her away from her house where her father had died recently. She was also on a regular follow-up with a psychiatrist for counselling. She delivered a full-term normal fetus of 3 kg and the puerperium was quite normal and there were no further manifestations of choreiform movements or nervousness of any kind. However, she is still seen on follow-up and is on penicillin prophylaxis to prevent rheumatic heart disease.

Table 1 Lab investigations of the patient

Haemoglobin	10.3 gm/dL
Total leucocyte count	7200/mm ³
ESR	10
C reactive protein	Negative
Anti-nuclear antibody	Negative
ASO titre	Negative
VDRL	Negative
Lupus anticoagulant	Negative
Serum calcium	8.0 mg/dL
Thyroid profile	Normal
Renal function test and liver function test	Normal
Urine routine examination	Normal
Throat swab culture	Negative

ASO, Anti-Streptolysin O; ESR, erythrocyte sedimentation rate; VDRL, venereal disease research laboratory; ASO, arteriosclerosis obliterans.

DISCUSSION

Chorea is an involuntary abnormal movement characterised by abrupt, brief, non-rhythmic and non-repetitive movement of any limb, often associated with non-patterned facial grimaces. The choreic movements may be caused by ischaemia or enhanced dopaminergic sensitivity mediated by increased female hormones during pregnancy. Chorea gravidarum is considered a syndrome rather than a specific disease entity. In recent times, most cases of chorea appearing during pregnancy in developed countries were caused by disease entities like systemic lupus erythematosus and Huntington disease. The decline in new cases of chorea gravidarum is probably the result of a decline in rheumatic fever which was a major cause prior to the era when antibiotics came into use for *Streptococcal* pharyngitis. However, the scenario is different in developing countries like India where rheumatic fever still occurs frequently. Rheumatic chorea also known as Sydenham's chorea is a delayed complication of group Aβ haemolytic streptococcal infections and forms one of the major criteria of acute rheumatic fever.¹ Sydenham's chorea is usually found in less than 10% of rheumatic fever patients. Group A streptococcal infection induces antibodies against the host neural tissues by a phenomenon called molecular mimicry.²

Acute chorea is a disease of childhood and over 80% of the cases occur between 5 and 15 years of age. The occurrence of the first attack after 15 years is uncommon except during pregnancy or related to the use of oral contraceptives.³ Of initial attacks, 80% occur during first pregnancies, and one-half starts during the first trimester.⁴ The present case also was a primigravida but chorea in this patient had manifested in third trimester following an episode of emotional stress following death of her father. Some 35% of patients with chorea gravidarum have a definite history of rheumatism and as expected, this patient had a positive history of having rheumatic fever at the age of 14 years. The patient's laboratory investigations did not show any evidence of recent streptococcal infection. As reported by Dilege *et al*,⁵ most cases of isolated Sydenham's chorea will have negative serology.

According to the case definition for rheumatic fever, documented evidence of previous streptococcal infection is found in only 20–30% of cases. Rheumatic chorea is one of the three circumstances in which the diagnosis of acute rheumatic fever can be made without strict adherence to the Jones criteria.⁶

The patient had mild form of chorea which was aggravated by stress of her kin's death and it subsided with one dose of long-acting penicillin, proper seclusion, rest, multivitamins and multimineral therapy. Patients with Sydenham's chorea are at risk for the development of rheumatic carditis particularly mitral stenosis and to prevent this, a regimen of daily penicillin prophylaxis should be instituted and maintained.⁶ Chorea in this patient did not affect her pregnancy and its outcome.

The present patient was a case of rheumatic chorea without carditis with previous history of rheumatism but she was at risk of developing rheumatic heart disease in future. Approximately 20% of patients who present with 'pure' chorea and are not put on secondary prophylaxis develop rheumatic heart disease within 20 years. Therefore, patients with chorea, even in the absence of other manifestations of rheumatic fever, require long-term antibiotic prophylaxis.⁷

Chorea in this case was helpful to the patient as she was started on penicillin prophylaxis which would prevent development of rheumatic heart disease in future.

Learning points

- ▶ Pregnancy is the time of changing hormonal milieu, it can lower the threshold for chorea and emotional outburst can precipitate it.
- ▶ Mild cases of chorea gravidarum can be successfully treated with hydration and isolation.
- ▶ Prompt diagnosis and treatment of chorea gravidarum gives good maternal and fetal outcome.

Contributors MP was the treating physician for the patient and performed detailed clinical examination. SU was the psychiatrist who was regularly advising the patient. AS is the obstetrician who carefully assisted in the patient delivering a healthy fetus. BSL compiled the case report and submitted it online.

Competing interests None.

Patient consent Obtained.

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