

Recurrent Chorea Gravidarum in Four Pregnancies

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ABSTRACT: An Arab woman developed recurrent chorea gravidarum in all four pregnancies that went to term, and none in the three pregnancies that ended in spontaneous abortion. She also developed an acute psychosis in one of these four pregnancies. A discussion of similar cases in the literature is undertaken.

RÉSUMÉ: Chorée gravidaire récurrente lors de quatre grossesses Nous rapportons le cas d'une femme arabe qui développe une chorée gravidaire à chacune de ses quatre grossesses à terme, mais à aucune des trois grossesses se terminant par avortement spontané. La patiente développa aussi une psychose aiguë lors d'une des grossesses à terme. Nous discutons les cas semblables dans la littérature.

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Chorea gravidarum is a rare condition and is becoming rarer. The case reported here illustrates many of the features of this disorder. In addition to the chorea, this patient developed an acute psychosis during one of her pregnancies, a complication of chorea gravidarum which has been reported previously.

CASE REPORT

In November 1979, a 21 year old, gravida 5 para 1, Arab woman presented with an acute behavioural change in her twentieth week of pregnancy. She was unable to give a history, but her mother and husband stated that one day she went into the bathroom quite normally but when she came out she was reluctant to talk, seemed withdrawn and dyspraxic, and indicated to them that she had seen something coming in through the bathroom window to attack her. She had heard earlier in the day that her husband intended taking a second wife — not an unusual phenomenon in this community.

On examination she was a healthy looking pregnant woman who adopted a peculiar curled posture in bed and avoided eye contact. She was alert but very uncommunicative, and through brief mono-syllabic responses indicated that she was oriented and was able to understand speech. She also managed to write her name and certain numbers on command.

Throughout the examination she was making frequent choreic movements with all four limbs and exhibited facial grimacing. There were no other abnormalities on neurological examination. The blood pressure was normal, there was no evidence of valvular heart disease, and the ECG was normal.

The following investigation were within normal limits or negative: urinalysis, complete blood count, sedimentation rate, blood sugar, urea, electrolytes, calcium, liver function tests, ceruloplasmin, VDRL, LE cells, anti-nuclear antibodies and ASO titre. An EEG showed infrequent intermittent irregular left fronto-temporal theta activity, but

a repeat EEG three weeks later was within normal limits. A CT scan done because of the findings on the first EEG was normal.

A psychiatric consultation was obtained, the opinion given being that the patient had acute catatonic schizophrenia, as opposed to hysterical dysphasia or voluntary refusal to talk. Her condition did not improve after her husband assured her that he had no intention of taking a second wife. Trifluoperazine 10 mg. tid was started, with gradual improvement of her psychosis. The chorea continued however, until she delivered a male infant, when it ceased dramatically.

Her family history was negative, but her past obstetric history was remarkable. In 1975 her first pregnancy ended in a spontaneous abortion at twelve weeks. In 1976 she gave birth to a full term male infant. During the second half of that pregnancy she exhibited similar chorea which ceased abruptly at the time of delivery. No medical advice was sought during that pregnancy. In 1977 and 1978, she had two further spontaneous abortions at twelve and sixteen weeks respectively, without developing chorea. Two further pregnancies took place in 1981 and 1982, each going to term and each associated with chorea gravidarum in the last trimester, ceasing dramatically after delivery. When last seen early in 1983 she was well with four healthy children.

DISCUSSION

According to Mulla (1958) the incidence of chorea gravidarum in the United States was 1 in 3500 pregnancies going to term. But the figures vary widely (Zegart and Schwartz, 1968), and in one survey, 113 out of 170 obstetricians replying to a questionnaire had never seen a case (Willson and Preece, 1932). Recurrent chorea gravidarum is even more rare. Out of 797 cases, 99 had recurrence of chorea in subsequent pregnancies (Willson and Preece, 1932). Out of these 99 cases, only 12 had four or more attacks. Using these figures, which admittedly go back to 1932, the case reported here would be expected to occur only once in

every 240,000 pregnancies. More recent statistics might suggest that a case like this is even more rare, because the incidence of chorea gravidarum and rheumatic fever has decreased dramatically (Agarwal and Foa, 1982).

This patient did not have chorea other than that in association with pregnancy; she did not have rheumatic fever and examination did not reveal evidence of heart disease. In the group of patients studied by Willson and Preece (1932) more than half reported an attack of chorea not associated with pregnancy, over one third had a history of rheumatic fever, and one third showed clinical evidence of heart disease.

In this patient, three pregnancies which ended in spontaneous abortion before 16 weeks of gestation were not associated with chorea, whereas the four pregnancies that reached term were all associated with chorea. Fleischman (1953) reported a 27 year old woman who had eleven pregnancies, six of which terminated early in spontaneous abortions without evidence of chorea, whereas the remaining five pregnancies were complicated by chorea in the last trimester. Chorea gravidarum starts in the first trimester in 50% of cases, in the second trimester in 35%, and in the third trimester in the remaining 15% (Zegart and Schwartz, 1968). In this patient the onset was in the second and third trimesters.

A prominent feature of this case was the psychiatric disturbance which developed during one of her pregnancies on the same day that she heard of her husband's impending marriage to another woman. This raised the question of an hysterical reaction, although she did not improve after assurances to the contrary. A psychogenic etiology for chorea gravidarum has been discussed by Weigner (1936). Amongst 950 cases of chorea gravidarum studied to determine the role of psychogenic factors, 137 cases seemed to show evidence of psychogenesis.

Fleischman (1953) reported that his patient developed choreic movements after attending the funeral of a relative. Willson and Preece (1932) stated that the acute psychoses were the most frequent complications occurring in 51 of their 797 cases, taking the form of mania, delirium and other psychoses which can become permanent. Beresford and Graham (1950) stated that the most important complication of chorea of pregnancy appeared to be the acute psychoses, and that a history of anxiety precipitating the attacks had been noted by many authors.

It is difficult to categorise the patient's speech disturbance, as there was no receptive or expressive dysphasia or dysarthria. However King (1950) reported that the psychoses complicate certain cases and that severe cases cannot even utter articulate sounds. McEllin et al (1948) described speech disturbances in all their five patients, describing these as inarticulate, unintelligible or slurred speech. Lewis and Parsons (1966) described dysarthria in all three of their patients.

The focal EEG slowing in our patient was of concern and led us to do a CT scan. In fact similar EEG abnormalities were noted previously by Zegart and Schwartz (1968) whose patient had "left temporo-occipital slow wave disturbance" which they attributed to drowsiness. Other authors who did perform EEG's on their patients described "an EEG which did not reveal any cortical discharges" (Fleischman, 1953), and "non-specific symmetric slowing of an EEG" (Agarwal and Foa, 1982). We could not find any reports of CT findings in this condition.

The rarity and very low mortality of chorea gravidarum undoubtedly account for the almost complete lack of pathological

data in this condition. Early reports describe congestion, edema and scattered petechial hemorrhages, as well as perivascular degenerative changes which are most severe in the corpus striatum, especially in the caudate nucleus (King, 1950). A recent neuropathological study by Ichikawa et al (1980) demonstrated nerve cell loss and astrocytosis within the striatum, especially in the caudate nucleus, which the authors believed accounted for the choreiform movements. Although chorea gravidarum is considered by many to be rare form of rheumatic (Sydenham's) chorea to which pregnancy predisposes (Beresford and Graham, 1950), it was interesting to see that in this study there was no clinical or pathological evidence, either within the brain or within other organ systems, to support a diagnosis of rheumatic fever.

It is not known why chorea occurs during pregnancy or with birth control pills (Fernando, 1966) or how hormonal steroids activate it (Donaldson, 1978). It is possible that a pre-existing neuropathological lesion may be activated by hormonal estrogenic effects (Barber et al 1976). It has recently been proposed that estrogen might act by changing the dynamic range of responsiveness of dopaminergic systems and thus may enhance the effects of pharmacological stimuli in either direction (Holsboer, 1982). Furthermore, Beresford and Graham (1950) described a woman who had chorea in the last three of her thirteen pregnancies in whom the chorea persisted after the last pregnancy. Chorea of Huntington's disease may also manifest itself for the first time during pregnancy (Bolt, 1968).

The chorea is self limiting in almost all cases, and often disappears dramatically in the first few days after childbirth (Donaldson, 1978). However several treatments have been advocated in the literature, varying from rest and seclusion (Willson and Preece, 1932) to intravenous and intramuscular sodium amytal (Fleischman, 1953), phenobarbital (Lewis and Parsons, 1966; Zegart and Schwartz, 1968), chlorpromazine and thiopropazate (Lewis and Parsons, 1966) and haloperidol (Patterson 1979). In treating the symptoms of chorea one should take into account the possible teratogenicity of the individual drugs, and the fact that this is a self limiting condition. Termination of pregnancy is considered only in exceptionally severe, progressive and resistant cases (Lewis and Parsons, 1966). Since chorea gravidarum is considered by many to be simply a rare form of rheumatic chorea to which pregnancy predisposes, penicillin prophylaxis during pregnancy and delivery is advocated by some to prevent rheumatic heart disease (Lewis and Parsons, 1966; Zegart and Schwartz, 1968).

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