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SHORT COMMUNICATION

RECURRENT HORMONE DEPENDENT CHOREA: EFFECTS OF OESTROGENS AND PROGESTOGENS

P. V. BARBER, A. G. ARNOLD AND G. EVANS

Department of Neurology, North Manchester General Hospital

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SUMMARY. A case of hemi-chorea is described in a young woman, initially following administration of an oral contraceptive pill and recurring in a subsequent early pregnancy. The chorea ceased with the removal of the aetiological factor on each occasion. Subsequent challenge with a combined oestrogen/progestogen pill resulted in recurrence of the chorea, but the patient has been successfully maintained on a progestogen without any further symptoms. Thus it would appear that oestrogen component was the precipitating factor for the chorea.

Chorea of pregnancy and chorea during oral contraception are well documented clinical entities. In the following case, recurrence of choreiform tremor was the first indication of pregnancy and an attempt was subsequently made to separate the effects of oestrogen and progesterone in its pathogenesis.

Case report

A 17-year-old girl was admitted for investigation in December 1973. Three weeks previously she had noticed difficulty in picking up small objects with the left hand. Following this, she developed jerky involuntary movements of the left hand, arm and leg, and began to drag her left foot. Shortly afterwards she developed twitching movements of the left side of the face and complained of slight dysarthria and dysphagia.

There was no definite history of rheumatic fever, but she had had 'growing pains' at the age of 11 years. At the time she was first seen, she had been taking the oral contraceptive 'Confer' (ethinyloestradiol 50 μg, norethisterone 1 mg) for 39 days.

On examination she was disabled by left-sided choreiform movements involving the arm, leg and face, of sufficient violence to be described as hemiballismus. There was slight left-sided hyper-reflexia, but no paresis or other focal neurological abnormality and no evidence of rheumatic carditis. Oral contraception was discontinued and treatment with chlorpromazine, 25 mg q.d.s. commenced with some immediate improvement. The involuntary movements gradually subsided over the next 4 weeks and she returned to work.

In August 1974 she was re-admitted following a recurrence of chorea of identical distribution and severity over the previous fortnight. She was not taking the pill, but, on inquiry,

Correspondence: Dr P. V. Barber, Department of Neurology, North Manchester General Hospital, Manchester.

her last menstrual period was found to have commenced 6 weeks previously and a pregnancy test was positive. Symptomatic treatment with chlorpromazine and tetrabenazine was less effective in controlling the chorea and termination was recommended on these grounds and the social circumstances of the pregnancy. Termination was carried out by suction evacuation. The chorea began to improve 4 days later and had almost disappeared by the fourth week.

It was clearly of some importance at this stage, in view of the patient's unmarried status and neurological history, to establish a safe and effective method of contraception. She refused to adopt another form of contraception and wished to continue with the pill if possible. In these circumstances it was considered justifiable to confirm firstly that her chorea was oestrogen 'dependent' and secondly to investigate the effects of progestogens. The combined preparation 'Confer' was initially re-introduced in January 1975 and produced a recurrence of chorea in the sixth week. Symptoms disappeared totally after withdrawal.

In March 1975, oral contraception was resumed with an oral progestogen, norethisterone acetate ('Noriday'). In the ensuing 8 months there has been no recurrence of symptoms and the patient is well, still taking this preparation, at the present time.

Discussion

Chorea during oral contraception was first described by Fernando in 1966. A further five cases were described by Lewis & Harrison (1969) and two further series have since been documented (Gamboa *et al.*, 1971; Riddoch *et al.*, 1971). In most cases the delay between commencement of the pill and the onset of symptoms has been between 3 and 6 months. In our case the latent period was remarkably constant for the three episodes at around 6 weeks. Hemi-chorea or chorea affecting predominantly one side of the body is the rule. In four of Riddoch's six cases (Riddoch *et al.*, 1971), symptoms began on the left side and continued to be unilateral, one left sided and the other right sided. All of Lewis's five cases (Lewis & Harrison, 1969) were predominantly unilateral and all but one gave a fairly clear history of antecedent rheumatic chorea. Fernando's original case (Fernando, 1966) was a right hemi-chorea with an antecedent history of rheumatic carditis. All the documented contraceptives have been oestrogen containing preparations. It has been considered that both in 'pill' chorea and chorea gravidarum a previous neurological lesion of rheumatic aetiology is reactivated by an altered hormonal environment, though nothing is known of the precise mechanism, and a previous history of Sydenham's chorea is by no means invariable. Chorea gravidarum has been reported as carrying a risk of mortality and morbidity to both mother and fetus, but the reported figures are extremely variable and must depend, amongst other things, on the violence of the chorea and its degree of response to treatment. It appears from the literature that, in general, the syndrome in modern times is both less frequent and less severe than previously, as are rheumatic fever itself and its other sequelae.

There are in the documented series a few other examples of chorea precipitated initially by oral contraception and recurring in a subsequent pregnancy. We have not been able to find a previous example of chorea associated with a progestogen preparation or indeed any instance of its use in such patients. The circumstantial evidence that oestrogens were instrumental in the pathogenesis of chorea is very strong in this case. The use of the more recently introduced progestogens in isolation as contraceptive agents has enabled us to postulate

that they may be free of the unwanted effects of oestrogens in this respect, though insufficient time has elapsed for an absolutely firm conclusion.

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