Hormone replacement therapy induced chorea

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Chorea gravidarum and chorea induced by the contraceptive pill are well recognised, albeit rare, in young women, who often also have a history of Sydenham's chorea in childhood. We report the case of a 57 year old woman who developed reversible chorea when prescribed hormone replacement therapy.

Case report

At the age of 13 years the woman contracted scarlet fever, closely followed by generalised Sydenham's chorea, lasting about six weeks. At around the time of her menarche she developed intermittent, classic migraine attacks with no temporal relation to her menstrual cycle. Her first two pregnancies were uneventful, but her third ended with a miscarriage at 13 weeks. There was no history of venous thrombosis, and she had never taken the contraceptive pill.

At 48 her migraine increased in severity and frequency. She was prescribed ergotamine tartrate, cyclizine hydrochloride, and caffeine hydrate (Migril), followed three months later by buclizine hydrochloride, paracetamol, and codeine phosphate (Migraleve). Two weeks later she developed right hemichorea, which reached a peak after six weeks and was considerably improved by the time she was admitted to hospital 10 weeks later. Computed tomography showed full blood count; thyroid and liver function tests; copper studies; measurements of erythrocyte sedimentation rate, antistreptolysin O titre, serum titres of autoantibodies including antinuclear factor, and electrolyte values; and all other investigations gave normal results except that thyroid microsomal antibody titre was 1:400. Syphilis serology gave negative results in blood and cerebrospinal fluid, which showed no abnormality. The chorea was thought to be secondary to consumption of cyclizine or buclizine, or both, and by discharge the following month it had almost disappeared. Her menopause occurred over the ensuing months.

In July 1989, aged 58, she was prescribed conjugated oestrogens and norgestrel for vaginal dryness. Within two weeks she again developed right hemichorea. Examination showed chorea of the right arm and leg and of the mouth, as before, with additional slight chorea of the left hand. Reinvestigation showed a normal full blood count, erythrocyte sedimentation rate, and creatine phosphokinase activity and no acanthocytes or autoantibodies, including antinuclear factor. Thyroid function was normal, but the thyroid microsomal antibody titre was 1:6400. Prothrombin time and partial thromboplastin time with kaolin were both normal. Lupus anticoagulant and anticardiolipin IgM and IgG were not detected. Computed tomo-ography showed mild cerebellar atrophy. Hormone replacement therapy was stopped and the chorea began to regress two weeks later, subsiding almost completely over the next four weeks.

Comment

Chorea induced by oral contraceptives,2 chorea gravidarum,3 Sydenham's chorea,1 and lupus chorea4 are intertwined in published reports. Many women developing chorea with oral contraceptives or chorea gravidarum have a history of alleged Sydenham's chorea. A few women initially diagnosed as having Sydenham's chorea, however, have lupus or the lupus anticoagulant syndrome. Moreover, chorea gravidarum may also occur in women with lupus.1

In our patient the initial episode of Sydenham's chorea, the onset of migraine, and the menarche were all temporally associated. Chorea did not occur during pregnancy but did recur in the same year as the menopause, which was associated with exacerbation of her migraine. The drug treatment was blamed for the recurrence of chorea, but the hormonal changes associated with the menopause might equally have been responsible.

The administration of hormone replacement therapy was causally related to the appearance of prominent chorea and its withdrawal to noticeable clinical improvement. Despite the history of migraine, one miscarriage, and chorea we have excluded lupus and the lupus anticoagulant syndrome as far as possible. She also had a lifelong tendency to develop mild fidgets at times of stress, and we have never seen her entirely free of involuntary movements. This raises the question of whether persistent rheumatic chorea exists.

To our knowledge there are no reports of chorea provoked by conjugated oestrogens with norgestrel. The prescription of hormone replacement therapy to postmenopausal women is growing. Doctors should be aware that such treatment might provoke chorea, especially in women with a history of Sydenham's chorea.

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