lesson of the month (2)

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Chorea

Two weeks after starting the oral contraceptive pill, a 16-year-old girl developed increasingly violent chorea and an evolving psychosis with prominent hallucinations, ideas of reference, and paranoia. An erythematous skin rash subsequently developed and Sydenham's chorea (SC) was diagnosed. Following neuroleptic medication and steroids, her chorea and psychosis subsided. This case illustrates that severe psychotic features can occur in SC. It is recommended that antistreptolysin O titres and antibasal ganglia antibodies are checked early in patients with evolving movement disorders and prominent neuropsychiatric features, as the window for modifying the course of this immune-mediated disorder may be narrow.

Lesson

A 16-year-old girl was admitted with an abrupt onset of generalised chorea associated with falls two weeks after starting Microgynon® 30. She was otherwise well, with no history of sore throat, ear infection, fever or arthralgia. On examination she was afebrile with generalised chorea, facial grimacing and milk-maid grip. She was anxious and displayed pressure of speech. General and cardiovascular examinations were normal. Despite stopping the pill, the patient's chorea persisted for the next 10 weeks, becoming so violent that she was confined to bed. She developed a pervasive psychosis with visual hallucinations and illusions (insects crawling out of electrical sockets and her fingers), auditory hallucinations (four to six male and female voices making abusive or critical comments in the second and third person) and tactile hallucinations (insects crawling over her hair and skin). She displayed ideas of reference and became increasingly more paranoid, feeling she was being watched and monitored. Suicidal ideation was expressed on several occasions. Five weeks after chorea onset, she developed a widespread erythematous macular skin eruption (Fig 1). Skin biopsy was inconclusive, and the rash subsequently improved.

Blood tests including renal and liver function, inflammatory markers, blood cultures, anticardiolipin antibodies and lupus anticoagulant were normal/negative. The antistreptolysin O titre (ASOT) and anti-DNAase B were raised at 800 iu/ml (n<200) and 480 units/ml (n<240) respectively 10 days after chorea onset suggestive of recent streptococcal infection. Antibasal ganglia antibodies (Institute of Neurology, London) showed strong positive binding to pyruvate kinase M1, with weak positive binding to neuron-specific enolase, supporting the presence of autoantibodies against basal ganglia. Magnetic resonance imaging brain scan with contrast, serial electrocardiogram and transthoracic echocardiograms were normal.

She was commenced on prophylactic dose antibiotics (clarithromycin as penicillin allergic). Neuroleptic medications including haloperidol, risperidone and amilsulpiride were tried and withdrawn due to unacceptable drowsiness. Five days of oral methylprednisolone produced no objective improvement. Quetiapine, titrated over four weeks was better toler-

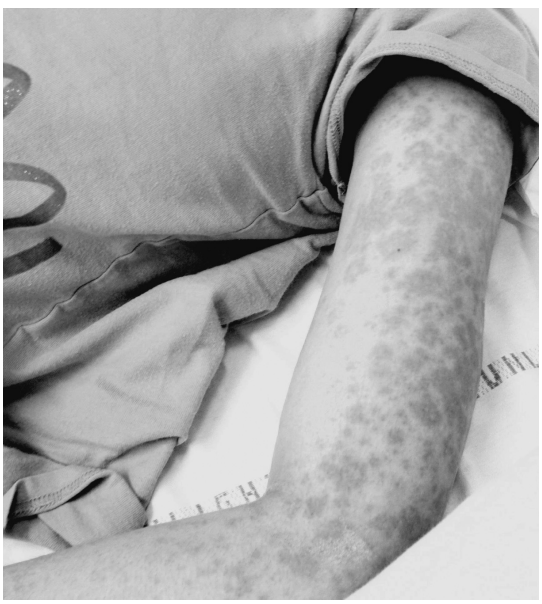

Fig 1. Photograph of widespread erythematous macular skin eruption on upper arm.

ated, with gradual reduction in the chorea and psychotic symptoms. Ten weeks after chorea onset, her involuntary movements and psychiatric features ceased, and she was discharged home on reducing quetiapine doses. One year later she remains well and off all medications, apart from prophylactic antibiotics.

Discussion

Since Thomas Sydenham's original description, Sydenham's chorea (SC) has become the prototype of a group of movement and neuropsychiatric disorders characterised by basal ganglia dysfunction due to an aberrant post-streptococcal autoimmune response and the presence of antibasal ganglia antibodies.[1] Common associated neuropsychiatriac features include anxiety, tics, altered behaviour/mood, obsessive-compulsive behaviour and attention-deficit hyperactivity disorder.[2] This case is unusual because of the severe psychotic features which were prominent throughout the duration of the choreiform illness and are rarely described in contemporary literature, although early similar case reports exist.[3] While relatively rare in the developed world, SC may be an under-recognised cause of an evolving movement disorder with neuropsychiatric features. The window for modifying the course of this immune-mediated disorder may be narrow, therefore early recognition and assay of ASOT/antibasal ganglia antibodies is crucial.

References

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