A recent case report by Konagaya and Konagaya in this journal noted the MRI findings in a subject with Sydenham's chorea, with abnormalities in the basal ganglia noted on a scan 31 days after the onset of the illness, but normal scans seen at day 45. In our similar case an MRI scan and a single photon emission computed tomography (SPECT) scan were normal.

A 22 year old woman presented with the acute onset of Sydenham's chorea. Three days before admission, she had noted paraesthesia of her left toe, and subsequently developed hemi-chorea of her left arm, face, and leg. Past history included mild asthma, atopic dermatitis, and mild iron deficiency anaemia as a result of menorrhagia. Medications included the oral contraceptive pill, inhaled bronchodilator, and an iron folate preparation. Examination confirmed the left hemi-chorea and hemiballismus with choreic movement of the tongue and face. She had a systolic murmur typical of mitral valve prolapse, but no signs of bacterial endocarditis.

Investigations included a normal full blood count, coagulation panel, and anti-nuclear antigen, β-human chorionic gonadotrophin, and thyroid function were negative or normal. A raised IgG cytomegalovirus antibody titre indicated past infection. The anti-DNAse B titre was elevated but antistreptolysin O titre was normal. Brainstem auditory evoked responses, CT, MRI, and SPECT scan of her brain were normal. Serum copper estimation was slightly elevated at 23-6 μmol/l (NR, 12–22 μmol/l), and her anticardiolipin antibody was positive. An EEG showed an excess of theta transients in the right central and parietal head regions. A transoesophageal echocardiogram was diagnostic of rheumatic valvular disease, showing thickening of valve leaflets associated with mild stenosis (valve area, 2 cm²; gradient, 4–5 mmHg). The left hemi-chorea persisted and oral tetrabenzine 25 mg twice daily was started with partial amelioration of the movement disorder. Penicillin 250 mg was started, and was, in fact, to continue until the age of 35 years. Advice was given about high dose antibiotic cover during dental or urogynaecological procedures.

The pathogenesis of Sydenham's chorea remains undefined. Its association with rheumatic heart disease was clarified in the mid-nineteenth century and the link between chorea and group A streptococcal pharyngeal infection was made 100 years later. Some anatomical studies of patients who had Sydenham's chorea note perivascular infiltrates in the basal ganglia and this supports a presumed ischaemic process. Cerebral imaging studies of subjects with Sydenham's chorea have usually been normal. One report in this journal of a patient with ballismus who had a cerebral MRI scan performed 31 days after the onset of chorea showed high signal intensities throughout the basal ganglia on T2 weighted imaging. These changes were not present on repeat MRI scan, and the authors considered the appearance to be consistent with neither ischaemia nor demyelination.

Pharmacological studies suggest an abnormal regulation of striatal dopamine, and dopamine depleting agents such as tetrabenazine have been used therapeutically. Sera from patients with Sydenham's chorea showed heterogeneous antineuronal antibodies, but the precise nature of these antibodies and target antigens is unknown. SPECT scanning is a technique that allows analysis of brain perfusion by using a radioactively labelled agent (exametazine), we scanned this patient's brain 10 days after the onset of chorea and showed a normal perfusion pattern. Together with a normal MRI 4 days after the onset of chorea, this case implies that there may be no acute perfusion abnormality in spite of post-mortem studies which often show basal ganglia vasculitis. This may either reflect the temporal profile of the condition or support the hypothesis that Sydenham's chorea is the result of an autoimmune process at the cellular level. Alternatively, the SPECT scanning technique may not be sufficiently sensitive to detect minor blood flow changes in deep cerebral structures.

A HILL
GK HERKES
Department of Neurology
P ROCHE
Department of Nuclear Medicine,
Royal North Shore Hospital,
Sydney, Australia


Paraneoplastic opsoclonus-myoclonus syndrome in metastatic ovarian carcinoma

Opsoclonus refers to involuntary, irregular, chaotic, conjugated eye movements in predominantly horizontal directions with a frequency of 6-12/second. Characteristically, an identical EEG pattern is lacking. Alternatively, the opsoclonus-myoclonus syndrome (OMS) may be an infrequent condition encountered as opsoclonus-myoclonus syndrome (OMS) when opsonoclonus is associated with focal or generalised myoclonus. Apart from rare causes such as vertebral ischaemia, haemorrhage of the pons or thalamus, hydrocephalus, coma, head injury, or the combined administration of haloperidol and lithium, OMS is commonly reported in viral encephalitis or as a remote manifestation of neoplasms. Although in children, the major paraneoplastic cause of OMS is neuroblastoma, in adults, carcinomas of the oat-cell type of the lung, melanoma, breast, and thymoma. Ovary is most frequently encountered. Recently, a young woman suffering from paraneoplastic OMS in Hodgkin's disease was reported in this journal. We describe OMS in the presence of ovarian epithelial ovarian cancer.

A 45 year old caucasian woman developed generalised shivers which severely interfered with walking two weeks before admission in November 1992. She also