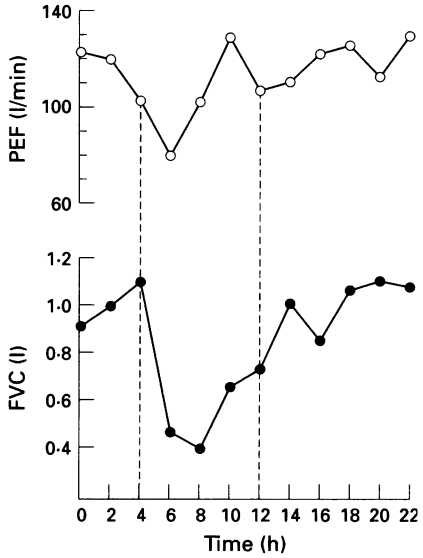

FVC and PEF plotted against time during the 24 hour period six days after the introduction of steroids. The time between the dotted lines represents a symptomatic decline in respiratory function.

next two weeks there was a steady improvement and the patient was discharged. Thefigure shows the FVC and PEF plotted against time during the 24 hour period six days after the introduction of steroids. The decline in respiratory function measured by the FVC was not matched by the PEF. The point that FVC measurements are needed to assess respiratory function in neuromuscular disease is well known but less than half of the dedicated medical neurology wards in teaching hospitals in England, Scotland, and Wales have a hand held spirometer capable of measuring FVC. A telephone audit of the neurology wards in teaching hospitals showed that 11 out of 24 units contacted (46%) possessed a spirometer or knew of one on a nearby ward. In London this figure fell to only three out of nine (30%). All neurological units should have their own spirometer.

C E G MOORE
E C WHEATLEY
M F JEFFERSON
D I SHEPHERD
Department of Neurology, North Manchester General Hospital, Delaunaya Road, Manchester M8 6RB, UK

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SPECT and MRI findings in Sydenham's chorea

A recent case report by Konagaya and Konagaya[1] 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 computerised (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 and biochemistry, and her antinuclear 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 cm^2; gradient, 4–5 mmHg) and mild regurgitation.

The left hemi-chorea persisted and oral tetrabenazine 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.[2] 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.[3] 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.[1] These changes were not present on repeat MRI scan, and the authors considered the appearance to be consistent with neither ischaemia nor demyelination.[1]

Pharmacological studies suggest an abnormal regulation of striatal dopamine, and dopamine depleting agents such as tetrabenazine have been used therapeutically.[4,5] Sera from patients with Sydenham's chorea showed heterogeneous antineural antibodies, but the precise nature of these antibodies and target antigens is unknown.

SPECT scanning is a technique that allows analysis of brain perfusion. 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

Correspondence to:
Dr G K Herkes, Department of Neurology, Royal North Shore Hospital, St Leonards, NSW 2065, Australia.

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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 intersaccadic interval is lacking.[1,2] An infrequent condition is encountered as opsoclonus–myoclonus syndrome (OMS) when opsoclonus is associated with focal or generalised myoclonus. Apart from rare causes such as vertebral ischaemia, haemorrhage of the pons or thalamus, hyperosmolar coma, head injury, or the combined administration of haloperidol and lithium, OMS has been reported in viral encephalitis or as a remote manifestation of neoplasms.[3,4] Although in children, the major paraneoplastic cause of OMS is neuroblastoma, in adults, carcinomas of the oat-cell type of the lung, uterus, breast, bladder, and thyroid gland are most frequently encountered.[3–5] Recently, a young woman suffering from paraneoplastic OMS in Hodgkin's disease was reported in this journal.[6] We describe OMS in the presence of metastatic 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

noted frequent involuntary movements of her eyes, heavily impairing object fixation. Family history was unremarkable. On admission, the patient was anxious, unconcentrated, and restless. She had opsoclonus with the conjugated eyes moving around arrhythmically in all directions with a large amplitude. Movements were also present with the eyes closed. Facial muscles showed diffuse myoclonic jerks, and there was myoclonic speech with dysarthria. No palatal myoclonus was seen; pupillary and the remaining cranial nerve functions were normal. There were no abnormalities in muscle tone and strength. Deep tendon reflexes were briskly elevated, the plantar sign being flexor. No sensory deficit was detected. Voluntary movements were impaired by frequent myoclonic jerking. The jerks increased to severe shivers, spreading out from the cervical region in cranial and caudal directions when the patient tried to sit upright, stand, or walk. Startling had the same effect. Truncal and limb ataxia and intention tremor were also observed. General physical examination revealed a deep tumour in the middle of the abdomen. There was atopic dermatitis of the face and the flexural areas of the extremities.

Laboratory investigations showed an increased ESR of 78 mm/h, a leucocytosis of 12×10^9/l, increased serum activities of γ-glutamyltranspeptidase (26 units/l) and lactic dehydrogenase (250 units/l). Other routine parameters were unremarkable. Carcinoembryonic antigen (5·0 ng/ml) and CA 125 (2161 units/ml) were elevated. Serum antineuronal antibodies anti-Yo, anti-Hu, and anti-Ri were not detected. Search for neurotoxic substances (thallium, neurotoxic drugs such as phenytoin, barbiturates, and benzodiazepines) in blood and urine was negative as were serum and CSF tests of bacterial, viral and fungal infections. CSF contained 3 cells, protein was 23·5 mg/dl. The Link–Tibbling index was 0·62. Isoelectric focusing of the immunoglobulins demonstrated oligoclonal bands in CSF, but not in serum. EEG was normal, without discharges time-locked to the myoclonic jerks. Electro-oculographic analysis (EOG) showed fast conjugated eye movements with amplitudes of 20–30° without intersaccadic interval in horizontal and vertical direction at a frequency of 8/s. MRI of the brain was normal. Further investigations included a radiograph of the chest, ultrasonography of the abdomen, CT scans of the chest and the abdomen, and total body bone scanning. A large cystic tumour was detected with presumed origin in the right ovary. Multiple enlarged lymph nodes were seen around the tumour, along the large vessels of the abdomen, behind the trachea, and in Virchow's node.

Laparotomy revealed a tumour of the left ovary of diameter 35 mm, with widespread lymphogenic metastases within the whole of the abdomen. Surgery was by supracervical abdominal hysterectomy, bilateral salpingo-oophorectomy, and infracolic omentectomy. Pathological diagnosis of the surgical specimens gave a solid ovarian carcinoma, moderately differentiated as an epithelial cell tumour with no oestrogen and progesterone receptors (International Federation of Gynecology and Obstetrics stage IIIc).

With this result, paraneoplastic OMS was diagnosed and treated by intravenous and oral clonazepam (up to 5 mg daily) and diazepam (up to 40 mg daily). The patient was subsequently able to stand and to walk a few steps with support. Two weeks after surgery, intravenous chemotherapy was started with carboplatin (350 mg/m²) and cyclophosphamide (600 mg/m²). Chemotherapy was repeated three times over the following five months, accompanied by a significant improvement in both ocular movements and myoclonic jerks. The patient was able to stand and to walk with aid, and the startle reaction was negative.

Although different aetiologies may cause OMS, its paraneoplastic form cannot be differentiated by signs and symptoms from other origins.[3] As in our case, paraneoplastic OMS usually precedes the detection of the underlying tumour by weeks or months. So far, carcinoma of the ovary is known to be associated with paraneoplastic neurological symptoms of acute or subacute cerebellar degeneration, sensorimotor polyneuropathy, and polymyositis.[7] From our case, this list may be supplemented by paraneoplastic OMS.

In paraneoplastic cerebellar degeneration, an autoimmune pathogenesis has been strongly suggested. Cases with ovarian adenocarcinoma show elevated titres of antibodies mostly against Purkinje cells ("anti-Yo-antibodies").[7] Other antibodies were directed against basket cells, stellate cells, and astrocytes. Antibodies against cerebellar cells were also detected in mixed mesodermal sarcoma of the ovary, oat-cell carcinoma of the lung, ductal carcinoma of the breast, adenocarcinoma and clear cell carcinoma of the uterus, colon carcinoma, and Hodgkin's disease. The antibody "anti-Hu" has been found in paraneoplastic encephalomyelitis together with small cell carcinoma of the lung, and in some cases of paraneoplastic cerebellar degeneration accompanied by encephalopathy and polyneuropathy.[7] The antibody "anti-Ri" directed against neuronal nuclei seems to be more closely related to opsoclonus. It has been found with malignant tumours of the breast, the fallopian tubes, and axillary lymph nodules when opsoclonus was evident clinically.[8 9] These observations may underline earlier suggestions that these antineuronal antibodies show some syndrome specificity. In our patient search for "anti-Yo", "anti-Hu", and "anti-Ri" antibodies were negative in serum. Whether this may be related to the different histological nature of the underlying ovarian tumour compared to those in the literature cannot, at present, be decided. Only oligoclonal banding in CSF gave a weak indication of an intrathecal immunological reaction.

No agreed treatment exists for OMS. Trials with adrenocorticotrophic hormone, steroids, or even plasmapheresis have not always been successful.[3 4 6 8] In our patient, clonazepam and diazepam reduced eye movements and myoclonic jerks substantially.[3–5] Paraneoplastic OMS has a slowly progressive course determined by successful treatment and the subsequent biological evolution of the underlying neoplasm.[3 5 7]

J SCHOLZ
P VIEREGGE
Department of Neurology,
C RUFF
Department of Gynecology and Obstetrics,
Medical University of Lübeck,
Germany
Correspondence to: Dr P Vieregge, Klinik für Neurologie, Medizinische Universität zu Lübeck, Ratzeburger Allee 160, D-23538 Lübeck, Germany.

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Cervical radiculopathy and bilateral internuclear ophthalmoplegia caused by temporal arteritis

A cervical radiculopathy and a bilateral classical internuclear ophthalmoplegia are rarely reported features of temporal arteritis.

A 57 year old man presented with a four day history of diplopia. During the preceding 10 days he had had right and then left sided facial and temporal pain. His medical history included untreated hypertension and mild asthma. He was a heavy smoker.

Examination revealed a painful pupil-sparing left third nerve palsy. Visual fields, fundi, and acuity were normal in both eyes. Tone and power were normal in the limbs and all reflexes were symmetrically present with flexor plantar responses. Blood pressure was 160/100 mmHg.

Blood investigations revealed a normal full blood count, normal urea and electrolytes, ESR 31 mm/hour, normal liver function tests, and a fasting plasma glucose of 7·1 mmol/l; a repeat fasting glucose was 5·6 mmol/l. A Venereal Disease Research Laboratory test was negative. A CT of the brain suggested the presence of an aneurysm of the ophthalmic artery on the left and revealed a pituitary tumour measuring 10 mm by 12 mm without suprasellar or lateral extension. Cerebral angiography confirmed the presence of a small broad based aneurysm at the origin of the left ophthalmic artery but no other abnormalities were found. Subsequent investigations showed that the pituitary tumour was secreting follicle stimulating hormone and that other aspects of pituitary function were normal.

Five weeks after presentation he developed weakness in both arms with discomfort around the neck and shoulders. He had also noticed some shortness of breath on exertion but did not complain of orthopnoea. Clinical examination revealed a residual left ptosis and a bilateral classical internuclear ophthalmoplegia with ataxic nystagmus in the abducting eye; convergence was preserved. In the upper limbs