LETTERS TO THE EDITOR

The moving ear syndrome: a focal dyskinesia

Although segmental dystonia of the cranial and upper limb muscles is well recognised, restricted and isolated dystonic movements of cranial involvement both such as those of the pinna are extremely uncommon. Dystonic movements of tranquil muscle groups such as “belly dancer’s dyskinesia” (dyskinesia of the abdominal wall), an axial tendon dystonia, and four cases of “moving ears” have been reported including two patients with unilateral involuntary twitching of the ear. We report a further two cases of unilateral movement disorder affecting the ear, one patient responding well to local injections of botulinum toxin.

Patient 1, a 23 year old white warehouseman complained of twitching of his right pinna since January 1994. Within three hours of development of the involuntary movement he experienced right temporal pain and a fluctuating noise in the left ear. There was no family history of any neurological disorder. The patient had no history of any serious illnesses in the past and was not on medication.

There was a continuous semimyotonic contraction of variable amplitude at a rate of 80 per minute, which both the ear and the scalp muscles above the ear. The involvement of the ear was more pronounced on the right. There was no palatal tremor or other dyskinesiae. Electromyography from the frontalis and auricularis superior muscles of the right ear, and the scalp muscles above the ear. The involvement of the ear was more pronounced on the right.

In patient 2, the movements are unlikely to be a form of tardive dyskinesia as the patient was aware of the movement disorder before starting neuroleptic drugs. Ten cases of “ear wigglers” due to tics of the ear were described by Keshavan. However, ear tic is unlikely in this patient as the movements were slow, rhythmic and not suggested by voluntary muscle contraction. In our patients, the slow often sinusuous movements of the ear with a superadded jerk element are suggestive of focal dystonia with myoclonic jerks. Also the presentation with a focal non-progressive movement disorder in adulthood is suggestive of dystonia. The reasonable responses to clonazepam in patient 1 and botulinum toxin injection in patient 2 suggest that the dystonic nature of these movements may be helped by standard treatment strategies for focal dystonia.

K RAY CHAUDHURI, P N LEIGH
W R G GIBB
The University Department of Neurology,
Institute of Psychiatry
King’s College School of Medicine,
London, UK
J F FYE
Department of Neurology,
Leicester Royal Infirmary,
Leicester, UK.

Correspondence to: Dr K Ray Chaudhuri,
University Department of Neurology, Institute of Psychiatry,
DeCrespigny Park, Denmark Hill,
London SE5 8AZ, UK.


Acute anterior horn cell disease resembling polioimmelitis as a manifestation of respiratory syncytial virus infection

Respiratory syncytial virus (Paramyxoviridae family) is an infectious agent of remarkable interest as it is the major cause of lower respiratory tract disease in young children. It can also cause infection in adults, although it is not so severe and does not have as much epidemiological importance as in infants.

Despite a high prevalence of respiratory syncytial virus infection, examples of neurological disease with a causal role have rarely been reported. Our patient developed an acute flaccid tetraplegia preceded by a meninigitic phase with serological evidence and positive cultures of a respiratory syncytial virus infection.

A previously healthy 28 year old man was admitted to hospital because of fever, meningism, and progressive weakness of the extremities. The patient had been vaccinated against polioimmelitis in 1966. A week before admission he developed an acute lower respiratory tract disease; four days later he began to have headache and diffuse weakness of all four limbs. The weakness was greater than distal. His 3 year old son had presented with a respiratory infection a week before the onset of the father’s symptoms. Examination showed a temperature of 38-7°C, signs of meningeal irritation, and proximal weakness of the limbs (grade 4-5).

Cranial nerves were intact. The tendon reflexes were hypoactive in both triceps and absent in the biceps and knees; the ankle jerks were normal. Plantar responses were flexor and no sensory abnormality was detected. His CSF had 70 white cells/ml (90% lymphocytes), 1.5 g/l protein, and 66 mg/dl glucose (103 mg/dl in serum). The second day in hospital he developed a progression of weakness with concomitant deterioration of respiration which required assisted ventilation. After 10 days sporadic fasciculations were noted. There were no extremities preceding the onset of a pronounced atrophy in all muscle groups and specially in the territory of C3 to C6 myotomes. Routine studies of blood and urine gave normal results. Tests for urinary porphobilinogen d-aminolevulinic, and anti-GM, ganglioside were negative. On the ninth day in hospital CSF examination showed 200 lymphocytes (95% lymphocytes), 3 g/l protein, and 77 mg/dl glucose. Antirespiratory syncytial virus antibody titre of 1/400 in serum and 1/1 in CSF were detected by direct immunofluorescence. Twenty five days later titre had increased to 1/1000 in serum and 1/10 in CSF. In addition the respiratory syncytial virus from CSF and bronchial aspirate samples was cultured in Vero and MRC-5 cell lines and identified using direct immunofluorescence (Monolux Screen RSV, Sanofi). The serological tests for other viruses and bacteria were negative. The patient had a history of alcoholic chronic pancreatitis and liver disease, but the alcohol abuse had been discontinued for 5 years. The autoantibodies were negative. The patient was treated with ribavirin (200 mg orally every...
### Neurophysiological results

<table>
<thead>
<tr>
<th>Motor</th>
<th>Day 3</th>
<th>Day 19</th>
<th>Day 90</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Radial cubital nerve</td>
<td>MAP (wrist/elbow) (mV)*</td>
<td>9±6/8 6</td>
<td>2/3</td>
<td>3±8-4</td>
</tr>
<tr>
<td>Delay latency (ms)</td>
<td>3-4</td>
<td>3-5</td>
<td>2</td>
<td>3-8±5</td>
</tr>
<tr>
<td>Conduction velocity (m/s)</td>
<td>57-9</td>
<td>58-9</td>
<td>8±4</td>
<td>8±4</td>
</tr>
<tr>
<td>F wave latency</td>
<td>3-7</td>
<td>3-7</td>
<td>3</td>
<td>3-7</td>
</tr>
<tr>
<td>Right ulnar nerve</td>
<td>MAP (ankle/popliteal fossa) (mV)</td>
<td>8±2-7</td>
<td>1±8</td>
<td>6±1</td>
</tr>
<tr>
<td>Delay latency (ms)</td>
<td>5-5</td>
<td>5-5</td>
<td>5</td>
<td>5-5</td>
</tr>
<tr>
<td>Conduction velocity (m/s)</td>
<td>53-2</td>
<td>53-2</td>
<td>53-2</td>
<td>53-2</td>
</tr>
<tr>
<td>F wave latency</td>
<td>58-1</td>
<td>58-1</td>
<td>58-1</td>
<td>58-1</td>
</tr>
<tr>
<td>Sensory:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right sural nerve</td>
<td>SAP (mV)</td>
<td>5-5</td>
<td>5-5</td>
<td>5-5</td>
</tr>
<tr>
<td>Conduction velocity (m/s)</td>
<td>52-9</td>
<td>52-9</td>
<td>52-9</td>
<td>52-9</td>
</tr>
</tbody>
</table>

*Muscle action potential recorded from the hypothenar eminence. †Muscle action potential recorded from the abductor hallucis. SAP = sensory action potential; NR = no response.

Eight hours for six weeks. This treatment did not improve the neurological damage but resulted in sterile CSF and bronchial cultures. We also used intravenous immunoglobulin and dexamethasone before laboratory data showed features of respiratory syncytial virus infection. Spine MRI was normal. Six months later serum immunoglobulin B, C, young, T, and NK lymphocyte subsets, CD4/CD8 ratio, lymphocyte blastogenic responses to both B and T mitogens, NBT test, and serum opsonisation capacity, and neutrophil and monocyte phagocytosis were normal. The patient began a slow recovery in the fifth week and artificial ventilation was discontinued on day 44. He started to walk unaided on day 70. After 10 months the patient was transferred to a rehabilitation centre. Motor examination at that time still showed grade 2-3 strength and there was considerable atrophy in the shoulder girdle muscles.

Neuropsychological studies were performed on the third, 19th, and 90th days after the onset of neuropsychiatric symptoms (table). The nerve conduction velocities, distal motor latencies, H reflexes, and sensory nerve action potentials were always normal. The first electromyographic examination showed slightly prolonged F wave latencies as a principal feature. Ulnar F wave response was absent on the next examination. The second and third studies showed a reduction in the size of the compound muscle action potentials. Electromyography showed mild to moderate denervation in the lower limbs and considerable denervation in the upper limbs, affecting mainly the C3-C6 innervated muscles. Values of somatosensory evoked potentials were in the normal range after median and posterior tibial stimulation.

Respiratory syncytial virus infection has rarely been associated with neurological abnormalities. In a few old reports respiratory syncytial virus infection could be serologically established in cases with polyradiculitis, meningitis or myelitis. More recently it has been documented in patients with Guillain-Barré syndrome and a case of Guillain-Barré syndrome preceded by a cold with serological evidence of respiratory syncytial virus infection has been documented. As far as we know, our case is unique in reports of respiratory syncytial virus associated neurological disorders for two reasons: the infectious agent could be cultured from CSF and the development of purely lower limb motor damage in a pattern similar to generalised polymyelitis. The major findings in this case were an acute generalised weakness and atomyroph without appreciable sensory change, spasticity, and, or clinical involvement of the corticospinal tract, and signs of meningeal infection. The EMG features place the likely site of the lesion at the anterior horn cell. The slowing of F waves in the first examination could be related to primary demyelination in the motor root but this feature can occur in motor neuron disease. The most effective chemotherapeutic management of respiratory syncytial virus infection is ribavirin. Our patient was treated with this antiviral agent after the 90th day; tetracyclasis was complete, but ribavirin treatment was effective in sterilising the CSF. In conclusion, respiratory syncytial virus infection should be incorporated in the differential diagnosis of the clinical syndrome of acute flaccid paralysis with meningitis. Because respiratory syncytial virus infection can be clearly diagnosed and is a treatable disorder, its recognition is important.

**Letters to the Editor**

J TEJADA
L E HERNÁNDEZ-ECHEBARRA
J F FERNÁNDEZ-LÓPEZ
Division of Neurology
JUÁREZ
Service of Clinical Neurophysiology
M F HERNÁNDEZ-NATAL
Service of Microbiology
D CARRIEDO
Medical Immunology Care Unit
J M GARCÍA
Immunology Unit
Complejo Hospitalario de León,
Calle Altos de Nava, sn,
24071 León, Spain.

Correspondence to: Dr Javier Tejada-García,
Servicio de Neurología del Complejo Hospitalario de León,
Calle Altos de Nava, sn,
24071 León, Spain.

Disorders of cobalamin (Cbl) are caused by inadequate intake, malabsorption, impairment of the extracorporeal transport, or inborn errors of intracellular Cbl metabolism.

1. Two Cbl-dependent enzymes are known: the adenosylcobalamin (AdoCbl)-dependent methylmalonyl-CoA mutase (MCM; EC 5.4.99.2) and the methylocobalamin (MeCbl)-dependent methionine synthase (MTH; EC 5.15.4-5.15.16). In the latter reaction, AdoCbl is oxidized to methylmalonyl-CoA (EC 2.1.1.13). Nine complementation classes of defects of intracellu-
lar Cbl metabolism or of the apoenzymes have been identified.

Hereditary defect of cobalamin metabolism (homocystinuria and methyl-malonic aciduria) of juvenile onset

Disorders of cobalamin (Cbl) are caused by inadequate intake, malabsorption, impairment of the extracorporeal transport, or inborn errors of intracellular Cbl metabolism.

1. Two Cbl-dependent enzymes are known: the adenosylcobalamin (AdoCbl)-dependent methylmalonyl-CoA mutase (MCM; EC 5.4.99.2) and the methylocobalamin (MeCbl)-dependent methionine synthase (MTH; EC 5.15.4-5.15.16). In the latter reaction, AdoCbl is oxidized to methylmalonyl-CoA (EC 2.1.1.13). Nine complementation classes of defects of intracellular Cbl metabolism or of the apoenzymes have been identified.