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 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 dyskinetia. Electromyography from the frontalis and auricularis muscles showed movements of movement and ear were presented. Our patient reported including dyskinetic and isolated dystonic movements of the ear were moved by the movements of focal dystonia. The reasonable reasons to clonazepam in patient 1 and 2. In the last 2 years old boy had presented with a respiratory infection at a week before the onset of the patient's symptoms. Examination showed a temperature of 38-39°C, signs of meningitis, and proximal weakness of the limbs (grade 4-5). Cranial nerves were intact. The tendon reflexes were hyperactive in both triceps and absent at the biceps and knees; the ankle jerks were normal. Plantar responses were extensor and no sensory abnormality was detected. His CSF had 70 white cells/mm³ (90% lymphocytes), 1.5 g/l protein, and 66 mg/dl glucose (103 mg/dl in serum). On 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 had increased and the patient presented with a worsening of the onset of the apparent atrophy in all muscle groups and especially in the territory of C3 to C6 myotonus. Routine studies of blood and urine gave normal results. Tests for urinary porphobilinogen δ-aminolevulinic, and anti-GM, ganglioside were negative. On the ninth day in hospital CSF examination showed leukocyte leucocytes/mm³, 3 g/l protein, and 77 mg/dl glucose. Antirespiratory syncytial virus antibody titres of 1/400 in serum and 1/1 in CSF were detected by direct immunofluorescence. Twenty five days later titres had increased to 1/1000 in serum and 1/10 in CSF. In addition the respiratory syncytial virus from CSF and bronchial aspirates samples was cultured in VERO and MRC-5 cell lines and identified using direct immunofluorescence (Monoflu Screen RSV, Sano). The serological tests for other viruses and bacteria were negative as was the blood culture. The respiratory symptoms and viral diseases were negative. The patient was treated with ribavirin (200 mg orally every

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

Respiratory syncytial virus (Paramyxoviridae family) is an infectious agent of remarkable interest because 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 relationship have been reported. 

Our patient developed an acute flaccid tetraplegia preceded by a meningeal phase with serological evidence and the presence 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 poliomielitis 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 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 38-39°C, signs of meningitis, and proximal weakness of the limbs (grade 4-5). Cranial nerves were intact. The tendon reflexes were hyperactive in both triceps and absent at the biceps and knees; the ankle jerks were normal. Plantar responses were extensor and no sensory abnormality was detected. His CSF had 70 white cells/mm³ (90% lymphocytes), 1.5 g/l protein, and 66 mg/dl glucose (103 mg/dl in serum). On 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 had increased and the patient presented with a worsening of the onset of the apparent atrophy in all muscle groups and especially in the territory of C3 to C6 myotonus. Routine studies of blood and urine gave normal results. Tests for urinary porphobilinogen δ-aminolevulinic, and anti-GM, ganglioside were negative. On the ninth day in hospital CSF examination showed leukocyte leucocytes/mm³, 3 g/l protein, and 77 mg/dl glucose. Antirespiratory syncytial virus antibody titres of 1/400 in serum and 1/1 in CSF were detected by direct immunofluorescence. Twenty five days later titres had increased to 1/1000 in serum and 1/10 in CSF. In addition the respiratory syncytial virus from CSF and bronchial aspirates samples was cultured in VERO and MRC-5 cell lines and identified using direct immunofluorescence (Monoflu Screen RSV, Sano). The serological tests for other viruses and bacteria were negative as was the blood culture. The respiratory symptoms and viral diseases were negative. The patient was treated with ribavirin (200 mg orally every

purely cultured from unique ratory by a meningoitis syncytial rarely muscles.

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 D in subclasses C3, C4, CH50, B, T, and NK lymphocyte subpopulations, CD4/CD8 ratio, lymphocyte blastogenic responses to both B and T mitogens, NBT test, T cell mitogen opsonisation competence, 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. Neurophysiological studies were performed on the third, 19th, and 90th days after the onset of neuropsychiatric 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 (GBS) 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 motor neuron damage in a pattern similar to generalised amyotrophy without appreciable sensory change, spasticity, 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 initial exacerbation 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 meningoitis. Because respiratory syncytial virus infection can be clearly diagnosed and is a treatable disorder, its recognition is important.

Letters to the Editor

Day 3 Day 19 Day 90 Normal
Motor: Right: cubital nerve
MAP (wrist/elbow) (mV)* 9.6/8.6 2.3 3.8/4.5 6.16
Distal latency (ms) 3.4 3.5 2.9 1.8-3.5
Conduction velocity (m/s) 57.3 58.4 49.5-71
F wave 34.7 NR 53.6 <31
Right: ulnar nerve
MAP (ankle/popliteal fossa) (mV)* 8.2/7.9 1.8/2 6.1/6.3 6.37
Distal latency (ms) 5 5.1 5 3.9-5.1
Conduction velocity (m/s) 53.2 50.3 45-60
F wave 58.1 49.1 47.2 <55
Sensory:
Right: sural nerve
SAP (uV) 5 5 2-19 1
Conduction velocity (m/s) 52.9 62.1 48-62

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

Neurophysiological results

Hereditary defect of cobalamin metabolism (homocystinuria and methylmalonic aciduria) of juvenile onset

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

Two Cbl-dependent enzymes are known: the adenosylcobalamin (AdoCbl)-dependent methylmalonyl-CoA mutase (MCM; EC 5.4.99.2) and the methylcobalamin (MeCbl)-dependent methionine synthase (MTHF reductase-methyl-Cbl-dependent methyltransferase: EC 2.1.1.13). Nine complementation classes of defects of intracellular Cbl metabolism or of the apoenzymes have been classified. The cblc/cblc and cblD/cblD is a rare autosomal recessive disorder reported in about 20 patients to date. Complementation analysis has disclosed two different complementation groups, CblC and cblD. Most patients present in early infancy with failure to thrive, developmental retardation, and megaloblastic anaemia. Manifestation later in life is rare. In one female patient the disorder manifested itself at age of 14 years as subacute myelopathy and dementia. After systemic OH-Cbl treatment cognitive improvement rapidly whereas the myelopathy responded slowly. In another patient the disorder manifested itself at age of 21 years and a diagnosis of multiple sclerosis was first made. After a six year relapsing and remitting disease course an isolated defect of methionine synthase (cblG) was detected.

Here a 30 year old woman (patient 1) with a 13 year disease course of relapsing and remitting myelopathy and neuropathy due to the cblc/cblc and her 34 year old sister (patient 2) are reported.

At the age of 12, patient 1 complained of unsteadiness of gait and urinary incontinence for a few weeks. Her motor and mental development had been normal. There was no family history of neurological disorders. She showed signs of a spinal cord disorder with pyramidal signs, and an impaired position sense. All ancillary tests, including vitamin B12 absorption and CSF studies, were normal. She had a complete remittance with prednisolone treatment. Two years later she developed similar signs, but more severe, which lasted several weeks until almost complete remission.

At the age of 19 another relapse occurred with first signs of a neuropathy with bilateral foot drop and absent ankle reflexes. Nerve conduction testing showed a reduction in amplitude and a low normal conduction velocity. A needle EMG showed chronic demyelination. Treatment with high dose corticosteroids resulted in partial improvement.

In the next two years the patient had an additional relapse occurred with progressive residual deficits and mild neuropsychiatric abnormalities. Cranial and spinal MRI and CSF studies were normal. A sural nerve biopsy showed a predominantly axonal neuropathy. During the next five months there was a progressive deterioration, with inability to walk, bladder incontinence, ascending sensory loss, severe respiratory alkalosis (pH 7.51, bicarbonate 34 mmol/l, base excess 10.9) with a severe disturbance of electrolytes (hypokalaemia (2 mmol/l), normal 3.5-5.5 mmol/l), hypomagnesaemia (0.8 mmol/l, normal 0.8-1.5 mmol/l)), and respiratory insufficiency. Serum Cbl was in the

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