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 musculature such as the muscles 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 torsion dystonia, and four cases of “shoving 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 fluttering 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/min involving all cranial and neck 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 muscles disclosed an absence of spontaneous activity and a normal CSF normal. A focal segmental dystonia involving the ear was diagnosed as part of the movement disorder in these patients merits discussion.

The movements, particularly in patient 1, had a jerky element, thus raising the possibility of segmental myoclonus and a relation to palatal tremor/myoclonus. Auricular myoclonus has been described in one patient, suggesting a central origin. Patient 1 had motor and sensory signs involving the frontalis bilaterally and this may occur rarely in palatal myoclonus. However, isolated ear movement as part of palatal myoclonus is unknown and neither of our patients had palatal myoclonus. The response of ear clicks. Furthermore, MRI in patient 1 excluded a brainstem lesion.

In patient 2, the movements are unlikely to be a form of tardive dyskinesia as the patient was aware of these movements 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 suppressed by voluntary muscle contraction. In our patients, the slow often sinusuous movements of the ear with a superadded jerky element are suggestive of focal dystonia with myoclonic jerks. The presentation with a focal non-progressive movement disorder in adulthood is suggestive of dystonia. The reason for centring 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.

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Acute anterior horn cell disease resembling poliomyelitis 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 relationship have rarely been reported. Our patient developed an acute flaccid tetraplegia preceded by a meningeal 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 poliomyelitis 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 38-7°C, signs of meningial irritation, and proximal weakness of the limbs (grade 4-5). Cranial nerves were intact. The tendon reflexes were hyperactive in both triceps and absent in the patellar reflex. The ankle jerks were normal. Plantar responses were flexor 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 serum). In the second day in hospital the patient developed a progression of weakness with concomitant deterioration of respiration which required assisted ventilation. After 10 days sporadic fasciculations were seen in the patient’s extremities preceding the onset of a pronounced atrophy in all muscle groups and especially in the territory of C3 to C6 myotomes. 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 200 leucocytes (95% monocytes), 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 immuno-fluorescence. 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 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 commonly associated with poliomyelitis and other viral disease were negative. The patient was treated with ribavirin (200 mg orally every
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