LETTERS TO THE EDITOR

The moving ear syndrome: a focal dystonia

Although segmental dystonia of the cranial and upper limb muscles is well recognised, restricted and isolated dystonic movements of cranial structures such as that 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 “moving ears” have been reported including two patients with unilateral involuntary twitching of the ear.1,2 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 semirhythmic contraction of variable amplitude at a rate of 80 beats per minute with both ears 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 showed normal motor unit firing in bursts. Brain MRI, CSF examination, and blood tests were normal except for an unexplained eosinophilia. The patient obtained considerable benefit from oral clonazepam.

The second patient, a 32 year old right handed man of West Indian extraction (born and raised in the United Kingdom) presented in April 1994 with a 12 year history of involuntary movement of the left ear. In 1983, he was thought to have a schizophrenic illness and was prescribed several forms of depot neuroleptic preparations (chlorpromazine, fluphenazine, flupenthixol decanoate) on which he has been maintained ever since.

The patient had been aware of abnormal movement of the ear at least a year before his schizophrenic illness and neuroleptic treatment. He had sought attention recently because the movements had become worse, were visible to others, and caused social embarrassment.

There was a slow rhythmic left ear elevation and retraction with occasional periods of posterior rotation and movement of the left temporals muscle. There was no evidence of any preceding or subjective symptoms and the movements could not be suppressed voluntarily. There were no abnormal movements of the right ear or of the temporalis, auricularis muscles or he did not have palatal tremor or any other dyskinesiae.

Routine blood tests, serum copper studies, and lysosomal enzyme screen were negative. Electromyography of the left auricularis superior and posterior muscles disclosed synchronous bursts of normal motor unit potentials lasting 200-300 ms at a frequency of 2 Hz. Recordings from the ipsilateral frontalis muscle and the right pinna were normal. Brain MRI was normal. Injection of botulinum toxin type A (40 mouse units, Dysport, Syntexicals Ltd) into the auricularis superior and posterior muscles gave appreciable benefit.

These two patients further illustrate the phenomenon of moving ears as a manifestation of focal dystonia. The nature of the movement disorder in these patients merits discussion. The movements, particularly in patient 1, had a jerk element, thus raising the possibility of segmental myoclonus and a relationship to palatal tremor/myoclonus. Auricular myoclonus has been described in one patient, suggesting a central origin.3 Patient 1 had movements of the right pinna face involving the frontalis bilaterally and this may occur rarely in palatal myoclonus.4 However, ear movements as part of palatal myoclonus is unknown and neither of our patients had clonus on myoclonus jerks on high pitched heard 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 the movement disorder before starting neuroleptic drugs. Ten cases of “ear wigglers” due to tics of the ear were described by Keshavan.5 However, ear tic is unlikely in this patient as the movements were slow, rhythmic and not suppressed voluntarily. We consider this to be a focal non-progressive movement disorder in adulthood in suggestive of dystonia. The reasonable responses to clonazepam in patient 1 and the complete lack of response in patient 2 suggest that the dystonic nature of these movements may be helped by standard treatment strategies for focal dystonia.

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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.1 Despite a high prevalence of respiratory syncytial virus infection, examples of neurologic disease with a causal role have been reported.2,3 Our patient developed an acute flaccid tetraplegia preceded by a meningeal phase with serological evidence and characteristic features 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 extremities. He was not able to walk or sit unsupported for more than six days. He was 3 years old but 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 hyporeactive in both triceps and absent in the patellar and ankle jerk. Deep tendon 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 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 were noted, with no further progression of the onset preceding the appearance of an atrophy only in muscle groups and specially in the territory of C5 to C6 myotonos. 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 leukocytes and meningitis (95% granulocytes), 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 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 (Monhoff Screen RSV, Sanofi). The serologic tests for other viruses and bacteria commonly associated with myelitis and encephalitis were negative. The patient was treated with ribavirin (200 mg orally every...