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Simultaneous occurrence of neuromyotonia and morphoea: a cause–effect relationship?

We report a case of unusual occurrence of neuromyotonia and morphoea (localised scleroderma) simultaneously in the same limbs. The patient was a 32-year-old man who presented with a 2-year history of twitching muscles in the right limbs with loss of hair and tightness of skin in the same limbs. Examination showed an almost complete loss of hair on the right arm, forearm and hand (fig 1). He also had hair loss on the right lower limb, especially on the back of the thigh and calf (fig 2). The skin in these areas seemed to be stretched and shiny with a parchment-like appearance. The other remarkable feature on examination was myokymia of the right-sided biceps, triceps, deltoid, brachioradialis, hamstrings and calf muscles. The rest of the neurological examination was normal. The clinical diagnosis was morphoea associated with neuromyotonia.

The antinuclear antibody was weakly positive. Anti-SCL-70 and anti-JO-1 antibodies were negative. CT scan of the chest was normal. Results of nerve conduction and repetitive nerve stimulation studies were normal. Examination by needle electromyography of the right deltoid, biceps, triceps and brachioradialis showed increased spontaneous activity, with single, doublet, triplet and long bursts of single motor unit potentials at rest (fig 3). Very long bursts (up to 60 s) of motor unit potentials akin to voluntary contraction were seen at rest in the right vastus lateralis. Needle electromyographic findings of the muscles of the left

Figure 1 Forearms and hands of the patient showing lack of hair on the right side.

Figure 2 Back of thighs and legs of the patient, showing lack of hair on the right side.

Figure 3 Needle electromyographic findings of the right deltoid showing motor unit potentials at rest.

limbs were normal. The result of skin biopsy from the affected area in the right arm showed a thinned out epidermis with hyperkeratinisation, with atrophic changes in the fibrous connective tissue in the dermis. Collagenisation and compression of fibrous tissue into dense compact collagen masses was seen in the dermis. Bundles of thickened nerves with round cell infiltrates were observed. These observations gave an impression of morphoea. A marked improvement in the neuromyotonia was noted within a fortnight of starting treatment with 300 mg/day phenytoin. No immunomodulatory treatment was considered because of the excellent response to phenytoin.

The most remarkable feature in our patient was the spatial and temporal coincidence of morphoea and neuromyotonia. This suggests an arcane relationship between the two conditions. The motor axon being the possible site of origin of neuromyotonia,^{1,2} one possible mechanism could be pathological change of nerves due to scleroderma.³ The appearance of nerve endings in the skin biopsy specimen lend credence to this view.

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Ear dyskinesia

Focal or segmental dyskinesias and dystonic syndromes affecting the cranial muscles are well recognised. Examples include the well known oro-bucco-facial dyskinesia caused by the long term use of neuroleptics, Meige syndrome, oromandibular dystonia, and blepharospasm. Among focal dyskinesias, ear dyskinesias are extremely uncommon and only a few cases have been reported.^{1,2}

Case report

We report a 57 year old woman who complained of involuntary movements of both ears for a year. The movements were initially intermittent but progressively became continuous. She could not control them voluntarily. The movements disappeared during sleep. There was no history of local trauma or neuroleptic treatment. Three months before onset, she had received paroxetine for depression. On examination there was a semirhythmic synchronous elevation and retraction of both ears. Movements were also seen in the skin around the ear. There was an intermittent contraction of frontal muscles as well. On clinical examination, there was amimia, and slow bilateral alternating serial movements. There were no tremor or hypertonias. MRI of the brain was normal. Electromyography of the left auricularis superior muscle showed bursts of normal motor unit potentials lasting 280 ms at a frequency of 2 Hz. The patient responded well to local injections of botulinum toxin type A (Botox) into the auricularis superior and posterior muscles (40 MU). The toxin was injected without EMG monitoring.

Discussion

The clinical appearance of our patient's movement disorder closely resembles dyskinesia because it consists of stereotyped focal or segmental movements affecting the ears and lasting for a few seconds or so. Focal dyskinesia can affect various regions of the body: ear, back, shoulder girdle and upper