Facial diplegia: cranial variant of Guillain–Barré syndrome?

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Simultaneous bilateral facial palsy is rare[1] and has various causes[2]. We report the case of a diabetic woman with transient severe facial diplegia as the principal neurological deficit.

CASE HISTORY

A Chinese woman aged 69 with a history of ischaemic heart disease with atrial fibrillation, hyperlipidaemia and recently diagnosed diabetes mellitus, reported sudden onset of drooling of saliva from both angles of her mouth and inability to smile. There was no complaint of difficulty in swallowing, visual disturbance, giddiness, limb weakness or numbness, or urinary or bowel symptoms. She was in atrial fibrillation with a heart rate 72/min, without postural hypotension. She was afebrile, and her neck was supple. On neurological examination there was severe bilateral facial weakness; she had lost her nasolabial fold bilaterally and was unable to frown, smile, blow, whistle or close her eyes tightly. There was no ptosis and extraocular movements were full. Both corneal reflexes were absent. Other cranial nerves functioned normally. Power in all limbs was grade 5/5, with normal reflexes except for hyporeflexic ankle jerks bilaterally. Her plantar responses were flexor. Sensory modalities and cerebellar function were normal. Blood counts and cultures, Venereal Disease Research Laboratory test and antinuclear antibody were negative; erythrocyte sedimentation rate, glycosylated haemoglobin and chest X-ray were within normal limits. Computed tomographic scanning of the head showed old lacunar infarcts in the right frontal region and left coronal radiata.

Tensilon test and repetitive stimulation tests were negative for myasthenia gravis. Blink reflex in both eyes showed absent ipsilateral R1 and R2 and contralateral R2 responses. On nerve conduction testing of the facial nerves there was bilateral prolongation of the latencies (right 4.5 ms, left 4.2 ms; normal <4.0) and amplitude (right 0.96 mv and left 1.18 mv; normal >1.5). Selective needle examination showed clear decreases in motor units in the frontalis and orbicularis oris bilaterally. No active denervations were seen. Nerve conduction studies of the peroneal and tibial nerves revealed a mild diffuse demyelinating polyneuropathy affecting the lower limbs (conduction velocities of motor nerves, peroneal right 35.9 m/s, left 35.1 m/s, tibial right 30.5 m/s, left 31.2 m/s left 31.2 m/s; normal >40). The F waves of the peroneal (right 58.7 ms left 51.5 ms; normal <50) and tibial (right 64.0 ms left 63.1 ms; normal <50) nerves were prolonged and the sensory superficial peroneal, sural nerves, and H reflexes were absent bilaterally. Needle sampling of selected muscles in all four limbs did not show any active denervations. Studies of the median and ulnar nerves were within normal limits. The patient declined cerebrospinal fluid examination. She was monitored for a week in the hospital without detection of new neurological signs.

Three weeks later, she turned up in our clinic with a broad smile. On clinical examination she had lost much of her facial weakness bilaterally. She was able to whistle, blow and frown. The corneal reflexes had returned to normal. There were no focal neurological deficits other than hyporeflexic ankle jerks. Repeat nerve conduction tests of her facial nerves showed improvement of her latencies (right 2.8 ms, left 2.8 ms) and amplitudes (right 1.4 mv, left 1.3 mv). Her blink reflexes had returned, with ispilateral R1 and R2 contralateral R2 all within normal limits. There was also improvement of the conduction velocities and F waves latencies of the peroneal and tibial motor nerves in the lower limbs (peroneal right 40.6 m/s, left 42.4 m/s; tibial right 35.2 m/s left 39.4 m/s; F waves peroneal right 53.3 ms left 50.0 ms; tibial right 54.4 ms left 54.0 ms). Her H reflexes, superficial peroneal and sural nerves, however, remained absent. Clinically, there was no objective sensory loss in the lower limbs.

COMMENT

Facial diplegia is present in less than 1% of patients with facial paralysis[1]. Reported aetiologies include inflammatory, infective, traumatic and infiltrative processes[2]. In addition to facial nerve involvement, one has to consider underlying neuromuscular and muscle diseases. Keane[2], in a 23-year review, found that out of 43 patients with bifacial palsy as the predominant sign, bilateral Bell's palsy (10/43) and Guillain–Barré syndrome (5/43) were the most common underlying causes. 2 of the 5 with Guillain–Barré syndrome progressed clinically; information on the others was not available. Other workers[3,4] have shown that facial diplegia was present in more than half the cases of Guillain–Barré

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syndrome, but the facial weakness was preceded or accompanied by limb weakness.

In our patient the weakness remained localized to the face with no clinical evidence of progression. The absence of blink reflexes (R1 and R2) indicated an abnormality in the trigemino-facial reflex arc on both sides. In addition, the abnormal facial nerve latencies and amplitudes suggested conduction blocks proximal to the site of stimulation of the facial nerves near their exit from the stylomastoid foramen. Needle examination of the facial muscles about a week from admission showed increased rate of firing of the neurons but no active denervations. This meant that there was no facial axonal degeneration. Nerve conduction tests of the peripheral nerves revealed a demyelinating neuropathy in the lower limbs with prolongation of the F waves and absent H reflexes. Repeat electrophysiological tests 3 weeks later showed return of the blink reflexes (both R1 and R2) and improvement of facial nerve latencies and amplitudes. This correlated well with the dramatic improvement of her bilateral facial weakness clinically. There was also improvement of the lower limb conduction velocities through the F waves. The persistently abnormal H reflexes and sensory nerve function may have been related to her diabetes mellitus.

Regrettably we could not persuade our patient to undergo a lumbar puncture for cerebrospinal fluid examination to document cytoprotein dissociation and confirm Guillain–Barré syndrome. But, taking into account the patient's clinical presentation, the dramatic improvement of her facial diplegia over a course of one month and evidence of a reversible demyelinating polyneuropathy on serial electrophysiological studies, we think she had a rare variant of the syndrome. While diabetes mellitus can be a cause for facial diplegia[2,5], the tempo of her clinical signs and progression and the good control of her diabetes suggested otherwise. Bilateral Bell's palsy can result in abnormal blink reflexes[5] but cannot explain the improvement in the conduction velocities and F waves in the lower limbs and the corneal reflexes. There was no significant travel or sexual history to suggest Lyme disease or human immunodeficiency virus (HIV) related infections.

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