Hemifacial spasm: treatment with pizotifen

The surgical treatment of hemifacial spasm has recently been reported in this Journal.1 I report the beneficial effects of pizotifen, a 5HT receptor antagonist, in two patients with hemifacial spasm. The original observation was entirely by chance as the drug was introduced to help a patient with frequent migraine attacks.

Patient 1, a 43 year woman pharmacist, had an eight month history of right hemifacial spasm after the removal of a melanoblast cyst from the lower eyelid in December 1994. There was also a background history of migraine headache fulfilling the criteria for the diagnosis as defined by The International Headache Society. Pizotifen (1 mg a day) was introduced by her general practitioner in an attempt to reduce the frequency of the migraine attacks. There was a rapid and consistent benefit from the point of view of the hemifacial spasm, although there seemed to be no reduction in the migraine attacks. The patient stopped the pizotifen and within 36 hours, the facial spasms returned. Several more trials of treatment, followed by stopping it, were undertaken, all with the same result. On continued pizotifen treatment, the benefit on her hemifacial spasm has been maintained.

Patient 2, a 52 year old woman clerk presented with a very long history of left hemifacial spasm. She had undergone various non-surgical treatments previously, including botulinum toxin, with variable benefit. There was also a background history of frontal headaches. These were more in keeping with a diagnosis of a tension type headache syndrome. In view of the previous case, pizotifen (1.5 mg at night), was introduced and there was very good improvement in the hemifacial spasm, although it was not completely abolished. The patient was considering a surgical approach for the spasm but decided in view of the considerable improvement that she would await events. The improvement has persisted for more than three months on continued pizotifen.

Considerable caution must obviously apply to the finding of just two patients benefiting from pizotifen treatment in hemifacial spasm. It is interesting, however, that the first patient, being a pharmacist, had undertaken several trials of stopping the treatment with the return of hemifacial spasm, only to see consistent improvement on restarting. The mechanism of action is unknown. Antagonising SHT1 receptors with pizotifen might inhibit neurons at the level of the Vth nerve nucleus.

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Orthostatic hypotension caused by a localised dorsal medullary tumour

Orthostatic hypotension is the most dramatic circulatory expression of autonomic failure. To control blood pressure, an intact baroreflex loop is necessary.1 In 1984, Hsu et al reported three cases of brainstem tumours that compromised baroreflex function.2 Here, we describe a patient who presented with severe orthostatic hypotension, respiratory failure, and bulbar paity associated with a medullary tumour. We examined the autonomic function of the patient. This case is unique, because of its rarity and diagnostic aspects.

A 54 year old man was admitted to Anjo Kosei Hospital on 21 April 1994 because of hoarseness, anorexia, dysphagia, and postural dizziness. These symptoms first appeared in March 1994. He had a history of ankylosing spondylitis and acute myocaridal infarction diagnosed at the ages of 26 and 51 years, respectively. His general examination was unremarkable except for ankylosis of the spine and a cachectic appearance. Neurological examination disclosed a nasal voice, atrophy of the tongue, and absence of the gag reflex bilaterally. Motor, sensory, and cerebellar functions were normal except for a mild proximal weakness of the limbs, probably due to disuse.

His gait was normal, and he could use a pair of chopsticks. The deep tendon reflexes were normal. There were no pyramidal signs. His blood pressure decreased from 160/100 to 80/60 mm Hg when he rose from a supine to a seated position, whereas his heart rate remained unchanged. He had no urinary dysfunction.

A plain cranial CT disclosed no apparent lesion. His CSF contained 38 cells per mm, and the protein concentration was 90 mg/dl. Repeated cytological analysis of the CSF disclosed no malignant cells. On 26 May, enhanced MRI showed a circumscribed brain tumour located in the dorsal medulla with involvement of the obex. Two additional smaller tumours also were found in the right cerebellum and in the right lateral periventricular (figure). No lesions in the spinal cord or peripheral nerves were noted. Nerve conduction studies of the limbs were normal. Repeated examinations of the whole body failed to disclose any evidence of a primary tumour. The intracranial tumour was considered to be a malignant lymphoma based on its radiosensitivity and neuroimaging.

After his admission, the patient's postural hypotension gradually worsened, and the frequency of syncopal attacks increased. A episode of respiratory arrest after syncope occurred during defecation on the morning of 7 June. He was unable to breathe spontaneously, and showed clouding of consciousness. However, he could respond voluntarily to some orders. Assisted ventilation was performed for the next two months. Irradiation therapy applied to his whole brain (40 Gy) and to the medullary lesion (14.4 Gy) was performed from 6 June to 19 July.

On 20 October, only small enhanced spots were seen on an MRI with contrast. His respiratory status improved and after 17 August, he was able to breathe without aid; however, his episodic orthostatic hypotension and bulbar deficits remained. Both sympathomimetic and mineralocorticoid agents given via gastric tube were ineffective in treating his orthostatic hypotension. The use of support stockings to prevent blood pooling in his

(A)

(Enhanced MRI (26 May 1994) shows a circumscribed brain tumour located in the dorsal medulla and involving the obex. (B) Enhanced MRI (20 October 1994) disclosing a significant decrease in tumour size after irradiation therapy. Only a few small enhanced lesions are visible.)
legs was beneficial, and he was able to walk occasionally without assistance.

Autonomic function tests were performed after weaning from mechanical ventilation. 1 A catheter was inserted into the left radial artery, and vital signs were monitored. Head up tiltting to 60° resulted in a blood pressure of 134/99 to 51/48 mm Hg. The basal plasma noradrenaline concentration was normal. A rise of plasma vasopressin in response to hypotension was absent, suggest- ing involvement of the afferent baroreflex pathway. There was no overshoot with Valsalva’s manoeuvre, which suggested insufficient baroreflex function. The absence of a blood pressure rise to the cold pressor test suggested involvement of the efferent sympathetic system. A noradrenaline infu- sion test disclosed sympathetically sensitiv- ity. No 1-vasopressin secretion in response to an isopropyl alcohol test. 1

Increase of his heart rate from 90 to 109 per minute in response to an atropine test showed that the vagal efferents were not completely involved. The results of the remaining tests are normal.

The control of blood pressure after pos- tural change is mediated by the baroreflex system. Failure of this reflex function can be due to a lesion anywhere in this reflex path- way, such as the baroreceptors, neuroregu- lating from the carotid sinus and aortic arch, travel via the glossopharyngeal and vagal nerves. Most of these baroreceptor afferents terminate in the nucleus tractus solitarius, the major neural relay of the baroreflex. 1, 2 In addition, the nucleus trac- tus solitarius not only regulates baroreceptor pathways but is also involved in the central regulation of respiration and swallowing. 3 The glossopharyngeal and vagal nerves also convey the sensory impulses from the pharyn-

x, larynx, and the respiratory system to the nucleus tractus solitarius, which contains specific centres for respiratory and swallow- ing. 

Our patient presented with severe orthostatic hypotension, dysphagia, lingual atrophy, and temporary failure of automatic respiration, but had no pyramidal signs, sen- soric symptoms, and aseptic meningitis, or a neur- ogenic bladder. Although we cannot discuss the relation of a non-metastatic autonomic neuropathy, neither spinal nor peripheral nerve lesions being responsible for the dysphagia, the neuritis exhibited in the dor- sal medulla may cause impairment of the nucleus tractus solitarius, the dorsal vagal nucleus, and the hypoglossal nucleus, which are involved in close proximity in the dorsal medulla. 4

In conclusion, a localised lesion of the dorsal medulla as reported in this case may result in severe orthostatic hypotension, bul- bar pseudoraphy which was an autonomic regula- tion in the absence of pyramidal or sensorimotor signs in the limbs.

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