LETTERS

Focal hand dystonia after cervical whiplash injury

There is currently a general consensus of agreement that dystonia is a disease of the basal ganglia, although dystonic symptoms have been observed in association with lesions in various different sites of the sensory and motor pathways.1 In particular, cervical intramedullary lesions have been reported as being a rare cause of focal hand dystonia, although in these cases the pathogenesis of the movement disorder remains unclear. To help clarify this point, we report the case of a patient who developed dystonic features of the right hand after a cervical whiplash injury.

Case report

A 44 year old man developed sensory alterations and impairment of strength in the right hand immediately after a whiplash injury. Neurological examination showed proprioceptive and tactile anaesthesia of the first three fingers of the right hand, while hyperreflexia on grasping, and adiadochokinesis of the right upper limb. Tendon reflexes, muscle tone, and plantar responses were normal, and thorough neurological examination of the upper and lower limbs also yielded normal findings. Cervical magnetic resonance imaging (MRI) revealed a small right posterior C5-C6 lesion of the spinal cord (see fig 1). Brain MRI, nerve conduction studies, EMG, and transcranial magnetic stimulation were all normal. Two months later, the patient developed writhing movements of the first three fingers and a dystonic posture of the right hand, worsened by movement, and more evident when the eyes were closed. Ability to write, use a knife or fork, and hold a glass were moderately impaired, especially without visual guidance. Neurological examination at this time revealed slight cutaneous and proprioceptive hypaesthesia and paraesthesias of the first three fingers of the right hand, while grasping strength was normal and the EMG recording showed a pattern of co-contraction of the forearm flexor and extensor muscles. Median nerve sensory evoked potentials (SEPs) revealed normal peripheral and central conduction times, but spinal and cortical waves were larger in response to stimulation of the median nerve on the dystonic side, and the spinal potential was slightly larger on the affected side. The asymmetry of SEP amplitudes was not attributable to different intensities of stimulation on the two sides, as the intensity of the peripheral shock was the same, as was the amplitude of the afferent volleys recorded at the elbow and at the Erb's point.

A larger N30 potential has previously been described in dystonic patients, but this finding has not been replicated in other studies.2 It may seem strange to find larger SEPs associated with a sensory deficit, but the presence of ephaptic spread in the lemniscal pathway may account for the larger afferent input to the cortex, as well as for the paraesthesias. We may postulate that this larger sensory input may have triggered an abnormal motor command resulting in movement disorder with features favouring dystonia over those seen with sensory deafferentation (that is, pseudoathetosis). This report may confirm the central role of abnormal sensory processing in the pathogenesis of dystonic symptoms.3

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References


Encephalomyeloradiculopathy associated with wasp sting

Although stings from wasps can cause severe allergic reactions, including anaphylaxis, neurological complications of wasp stings are rare. There are, however, various interesting case reports of acute myelitis, acute encephalitis, encephalomyeloneuropathy, optic neuritis,3 cerebral infarction,4 and acute inflammatory polyradiculopathy.5 We report here the case of a young man who developed encephalomyeloradiculopathy after being stung by a wasp.

Case report

An 18 year old man was referred to Ramathibodi Hospital with impaired consciousness and quadriplegia. He had been stung by a wasp 16 days previously on the right cheek and had mild swelling and tenderness over this area. On the following day, he suffered from headache, fever, and nausea and was admitted to a regional hospital. Three days later, he was drowsy and had urinary retention. Then he developed a generalised tonic-clonic seizure lasting two to three minutes. He was intubated and referred to us.

At Ramathibodi Hospital, he was comatose, quadriplegic, and areflexic. A lumbar puncture was done. The CSF pressure was 360 mm H2O and fluid analysis showed mononuclear cells (9/mm3), a protein concentration of 160 mg/dl, and CSF/blood sugar concentrations of 4.38/13.38 mmol/l. CSF and serum were tested for

Figure 1 Cervical T2 sagittal section (A) and axial section at the level of the C5-C6 intervertebral space (B). The arrows indicate the small right posterior lesion.
Hymenoptera urinary and fecal incontinence. The wasp was admitted, he was discharged with residual performed. The power of the upper extremities in developing both tibial and peroneal nerves. Sensory nerve conduction was normal in median, ulnar, and sural nerves. Methylprednisolone was given intravenously for five days. On the sixth day after starting treatment, he regained consciousness with limited eye movement and quadriparesis. A month later, a plasma exchange was performed. The power of the upper extremities gradually improved. Three months after admission, he was discharged with residual paraplegia with sensory level at T1 and urinary and fecal incontinence. The wasp was identified as Vespa tropicalis, a wasp commonly found throughout the country.

Comment
Allergic reactions to Hymenoptera stings range from local to severe systemic reactions or even death. These reactions are usually acute, beginning within minutes to hours in 76–96% of the patients. Nevertheless, there are reports of delayed reactions that can occur days to weeks after the event. Of the 2606 reactions noted in the 1964 Academy of Allergy survey, 2.8% did not occur until several days after the sting. There have also been reports of neurological complications, hyperglobulinaemia, thrombocytopenic purpura, n Ephthic syn- drome, and hepatoportal syndrome. The neurological complications are infrequent but often serious and include clinical manifestations of damage to the central and peripheral nervous system.

Means et al reported a case with a relapsing and progressive course of neurological symp- toms and signs, including bilateral weakness and numbness of the arms and legs, following a sting by a yellow jacket (Vespa pensylvanica). This patient had been alert and oriented throughout her clinical course but she eventually died after sudden respira- tory and cardiac arrest. Necropsy revealed massive pulmonary embolism which was the cause of death. Examination of the nervous system showed areas of demyelination throughout the central and peripheral nerv- ous system associated with necrosis and inflammatory infiltrates in the brain stem and spinal cord. This is the only previous report of encephalomyelopolyradiculoneuritis in the English language literature. In contrast to this, our patient had a more fulminating clinical course with a seizure and alteration of consciousness. It appears that there are occa- sional reports of optic necephalitis, and encephalomyelopolyradiculoneuritis in the Russian and Romanian literature but the abstracts of those reports were not available for review.

Maltzman et al reported two cases and reviewed five other cases of optic neuropathy after bee and wasp stings. Most cases had significant visual recovery after corticosteroid treatment. Bachman et al reported five cases presenting with acute inflammatory polyra- diculopathy following Hymenoptera stings, with good recovery. Some cases had a sural nerve biopsy which showed segmental demy- elination.

From early reports and our case, it appears that patients with neurological complications after Hymenoptera sting usually improve and some have complete recovery after high dose corticosteroid treatment. According to some necropsy reports, the course of the disease, and the response to treatment, the pathogenesis—although not definitely known—could be an inflammatory response to Hymenoptera sting. The nature and location of the sensitising agents involved in Hymenoptera are not entirely clear. The venom, venom sack, and insect body have all been shown to possess antigenic properties. Hymenoptera venom contains various non-myelin proteins or peptides that could be encephalitogenic in some individuals. The antigens of the wasp may initiate production of antibodies that would cross react with myelin basic proteins. Alternatively, the phospholipase A activity of the venom could liberate encephalitogenic basic proteins (including from myelin) from membranes of the central and peripheral nervous systems, inducing immune reactions. Although definitive evidence of the relation between an envenoming sting and neurological complications is often missing, this is true of many conditions that reflect delayed immune responses. It is hoped that a more detailed systematic evaluation of the consequences of severe stings by Hymenoptera, including serological and immunological testing, will solve these problems in the future.

Figure 1 Magnetic resonance imaging (T2 weighted) of brain and cervical cord showing multiple ill defined scattered lesions of inhomogeneous isohyperintense signal involving both grey and white matter of medulla, pons, midbrain, basal ganglia, thalami, centrum semiovale, cortical grey matter, and cervical cord.

Debrisosquino hydroxylase gene polymorphism (CYP2D6*4) in dementia with Lewy bodies

After Alzheimer’s disease, dementia with Lewy bodies (DBL) is probably the second most common cause of dementia among the elderly, having been shown to account for around 20% of cases at necropsy. Pathologi- cally, DBL is characterised by the presence of Lewy bodies within the cerebral cortex, espe- cially the parahippocampal gyrus, cingulate gyrus, and temporal neocortex. Some genetic association studies, and within brain stem nuclei, principally the sub- stantia nigra and locus caeruleus. Nonetheless, histopathological changes classically associ- ated with Alzheimer’s disease (amyloid plaques and neurofibrillary tangles) are fre- quently widespread within the cerebral cortex of patients with DBL.

Although most cases of DBL appear to arise sporadically, cases with a previous family his- tory of similar disorder are known, suggesting that genetic factors may contribute to the risk of developing disease. It is well recognised that cases of DBL, especially male, show an increased frequency of APO E epsilon4 allele, although possession of this is generally associ- ated with an accompanying Alzheimer’s dis- ease type pathology, with DBL cases without Alzheimer’s disease type pathology having a normal frequency of APO E epsilon4 allele frequency (Bachman et al, unpublished data). Hence, possession of APO E epsilon4 allele per se is unlikely to contribute to the generation of the Lewy body component of the pathological spectrum.

Some genetic association studies in idiopathic Parkinson’s disease have reported an increased frequency of the CYP2D6*4 allele of the debrisoquino hydroxylase gene (involved in the G/G transition at the intron 3-exon 4 junction) which results in a inactive copy of the enzyme and a “poor metaboliser” pheno- type. Other work has suggested that this same allelic variation may also occur more fre- quently in DLB, but not all studies agree.

We have examined the frequency of the CYP2D6*4 allele of the debrisoquin hydroxy- lase (DBH) gene in 53 patients with DBL. The clinical diagnosis of DBL was made in accord- ance with the consensus criteria of McKhirt et al. Twenty five of the patients have died, and pathological examination of their brains (DMAM) confirmed the clinical diagnosis in every instance. Genomic DNA was extracted from blood (in living patients) or frozen brain tissue (in necropsy cases) by standard methods. DBH and APO E and genotyping were performed according to standard methods.

Differences in APO E epsilon4 allele and DBH CYP2D6*4 allele frequency between patient and control groups were analysed by Fisher’s exact test. As previously reported, the APO E epsilon4 allele frequency was significantly increased (compared to controls) in both pathological and pathologically DBL groups, separately or com- bined (table 1). However, in agreement with previous reports, there were no significant differences in frequency of CYP2D6*4 allele of DBH gene between DLB cases (clinical or pathological groups (separately or combined).
and control subjects. Neither were there any differences in age at onset of disease or (in the pathological cases) duration of illness between DLB cases with and without mutant CYP2D6*4 allele. Cases with CYP2D6*4 allele were no more likely to show any, or more severe, Alzheimer’s disease type pathological changes than those without. Hence, in this present series of cases of DLB we have not been able to confirm possession of CYP2D6*4 allele in the pathogenesis of the disorder, either in terms of generating Alzheimer’s disease or Lewy body type pathology or in influencing the age at onset or duration of the illness. We therefore conclude that possession of CYP2D6*4 allele of DBH gene does not act as a risk factor for DLB.

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References

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Meningioma presenting as stroke: report of two cases and estimation of incidence

Meningioma is the most common extra-axial brain tumour in adults and frequently originates in the suprasellar, frontobasal, temporo-basal, sphenoid wing, or petroclinoidal regions. Tumours situated in these locations often involve an intracranial portion of the internal carotid artery (ICA) and may compromise cerebral blood flow. While transient cerebral ischaemia has been recognised as a complication of skull base meningiomas, to our knowledge, there are no documented cases of a meningioma causing stroke by ICA occlusion. We report two cases of meningioma presenting with cerebral infarction as a result of carotid artery compression and estimate the incidence of meningioma related cerebral ischaemia by this mechanism at our institution.

Case reports

Patient 1
A 49 year old right handed man experienced two weeks of left upper extremity weakness. Brain magnetic resonance imaging (MRI) reportedly revealed a right sided mass involving the cavernous sinus, and the patient was referred for a neurosurgical consultation. Two days later the patient noted a sudden increase in symptom severity and presented to an outside hospital. Neurological examination showed a left facial droop, left hand weakness of 4/5, and decreased sensation on the left side of the face.

Brain MRI demonstrated a subacute right middle cerebral artery (MCA) territory infarct. In addition, there was a mass in the right cavernous sinus, consistent with a meningioma, which encased the ICA (fig 1A). Cerebral angiography demonstrated occlusion of the right cavernous ICA with radiographic blush of the right orbit (fig 1C). The mass also compressed the right optic nerve. Cerebral angiography demonstrated near occlusion of the right distal ICA (fig 1D) with the majority of perfusion to the right hemisphere being supplied by cross filling from the left ICA. Transosseous vaginal echo-cardiography was normal without evidence of embolic source. Neuro-ophthalmological examination revealed no signs of emboli. A comprehensive serum hypercoagulable panel revealed no abnormalities.

The patient underwent focused radiation therapy of the meningioma and his symptoms gradually improved. However, about six months later he experienced worsening left leg weakness. Brain MRI revealed an acute right MCA and right watershed distribution infarct. There appeared to be fresh thrombus in the right cavernous ICA. The patient was given anticoagulation and had no further neurological events in over 30 months of follow-up.

Patient 2
A 31 year old right handed man had acute onset of complete visual loss in his right eye. The event was painless, and as the patient felt that he could compensate sufficiently, he did not seek medical attention. One morning, about two years later, he experienced sudden onset of left sided numbness, collapsed, and was unable to sit up. Brain MRI revealed a right parietal region infarct, as well as a large mass, consistent with a meningioma, originating from the olfactory groove and encasing the ICA at the apex of the right orbit (fig 1E). The mass also compressed the right optic nerve. Cerebral angiography demonstrated near occlusion of the right distal ICA (fig 1F) with the majority of perfusion to the right hemisphere being supplied by cross filling from the left ICA. Transosseous vaginal echo-cardiography was normal without evidence of embolic source. Neuro-ophthalmological examination revealed no signs of emboli. A comprehensive serum hypercoagulable panel revealed no abnormalities. The patient underwent complete tumour resection. Over the next several weeks he recovered most neurological function but was left with no vision in the right eye and persistent left arm numbness.

Discussion

Meningiomas are prevalent brain tumours commonly located at the skull base. By virtue of their position, these tumours have the potential to affect portions of the ICA and compromise cerebral blood flow. Previous reports have suggested that meningioma related compression of the carotid artery may produce transient neurological symptoms including loss of consciousness, hemiparesis, paraesthesia, and global amnesia. To our knowledge, however, there has never been a documented case of cerebral infarction as a result of meningioma related ICA compression.

We present two patients with cerebral infarction attributable to meningioma related ICA involvement. In case one, imaging after the first ischaemic episode demonstrated a large cavernous sinus meningioma surrounding and occluding the right ICA. MRI after the second ischaemic episode suggested stump thrombosis in the cavernous ICA. The stroke was probably attributable to both haemodynamic hyperperfusion as well as artery to artery embolisation. In case two, imaging demonstrated a large ollactory groove meningioma encasing the right ICA with near occlusion of the vessel. It seems his stroke also occurred because of a combination of hyperperfusion and thromboembolism. Neither of these patients had evidence of vasculopathy or another aetiology for stroke.
Although meningiomas commonly involve the ICA, they rarely present with symptoms of cerebral ischaemia. These tumours typically do not change vascular patency even when completely encasing the ICA and its bifurcation into the MCA and anterior cerebral artery. It may be possible that meningiomas, being slow growing and non-invasive, do not exert sufficient external force to significantly compress the high pressure arterial vasculature. In addition, the ICA vessel wall is thick with a muscular media segment, thereby offering substantial resistance to vascular compromise. By comparison, cortical veins and dural sinuses, being low pressure compartments with thin walls, are frequently compromised by meningiomas. The tumour’s slow growth rate, however, allows for development of substantial collateral drainage, and as a result, cortical infarction attributable to venous insufficiency has only been reported postoperatively after injury to these compensatory pathways.

The rate at which meningiomas present with symptoms of cerebral ischaemia is unknown. In an attempt to estimate the incidence at which meningiomas manifest symptoms of cerebral ischaemia by ICA compression, we reviewed retrospectively the medical records of 1617 patients with meningiomas evaluated by the surgical neuropathology service at our institution from 1985 to 2001. We identified three patients with menigiomas manifesting the ICA compression. The first patient had a parasellar/medial sphenoid wing meningioma that narrowed the right ICA within the cavernous sinuses and presented with progressive left hemiparesis. The second patient had a petroclival meningioma that encased the left ICA and presented with right upper extremity paresthesias. The third patient had a frontobasal meningioma that involved the right ICA and presented with evolving left hemiparesis. Thus, while meningiomas frequently involve intracranial portions of the ICA, we estimate the incidence of meningioma related cerebral ischaemia by carotid artery compression to be only 3 of 1617 tumours or 0.19%.

In conclusion, we describe two cases of cerebral infarction as a result of carotid artery compression by a meningioma. We hypothesise that meningiomas typically do not compromise the ICA significantly because of the slow growth rate and non-invasive nature of the tumour, as well as the high arterial pressures of the ICA. Consequently, cerebral vascular insufficiency is an exceedingly uncommon presentation for meningioma.

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The relation between daytime sleepiness, fatigue, and reduced motivation in patients with adult onset myotonic dystrophy

Daytime sleepiness, apathy, and lack of motivation are established clinical manifestations of myotonic dystrophy. A recent study showed that modafinil reduced daytime sleepiness and average sleep latency in a group of nine patients with myotonic dystrophy. This finding suggests that daytime sleepiness in patients with myotonic dystrophy and without obstructive sleep apnoea might be central in origin. A magnetic resonance imaging study indeed found evidence for a possible association between cerebral abnormalities in myotonic dystrophy and excessive daytime sleepiness.

Although several lines of evidence suggest that dopamine release in the striatum is regulated by serotonin, the effects of serotonin and SSRIs on dopamine release in the striatum of normal animals are disputed. Some studies have demonstrated that stimulation of the 5-HT(1A) receptors inhibits dopamine release and increases tyrosine hydroxylation in the striatum. In the striatum of the animals with nigrostriatal dopaminergic denervation, 5-HT(1A) receptor density was upregulated. The density of dopamine D2 receptors in the striatum was increased after repeated administration of milnacipran. Infarcts in the basal ganglia might have impaired such adaptive changes in the dopaminergic system, rendering the patient susceptible to milnacipran induced parkinsonism.

To my knowledge, this is the first reported case of parkinsonism associated with the use of SNRI. Clinicians should be aware that not only SSRIs but SNRIs can cause severe parkinsonism.

Competing interests: none declared.

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References

In the light of these results we would like to advocate the simultaneous use of both daytime sleepiness and fatigue outcome measures in future treatment and fatigue studies.

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Parkinsonism associated with a serotonin and noradrenaline reuptake inhibitor, milnacipran

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