

circulation as the mechanism of the spinal cord injury.

Given the lack of effective medical intervention for this condition, the emphasis should be on prevention. In the cases associated with operative positioning, use of intra-operative neurophysiological monitoring would be advocated and argued to prevent this complication by detecting early evidence of spinal cord dysfunction during the procedure,<sup>4</sup> and in the cases of substance abuse raising community awareness of the devastating long-term irreversible neurological compromise and quadriplegia is imperative.

#### CONCLUSIONS

We believe that Cervical Flexion Myelopathy is an under-recognized clinical diagnosis and reporting such cases is important to raise awareness of the possibility of this complication, with the goal to improve or increase preventative measures.

*Hussein Alahmadi, Gelareh Zadeh  
Toronto, Ontario, Canada*

#### TO THE EDITOR

##### Isolated Unilateral Hypoglossal Nerve Palsy

Hypoglossal nerve palsy is a rare cranial neuropathy with a broad differential diagnosis. Most etiologies typically present with other neurological or systemic sequelae(1). Thus, isolated hypoglossal nerve palsy presents a diagnostic challenge requiring a systematic investigative approach. We report of an interesting case of isolated unilateral hypoglossal nerve palsy following a mononucleosis infection. The investigative work-up, differential diagnosis, and management are discussed.

A 57 year-old male presented with a 6-month history of a swollen left tongue, decreased tongue mobility, and dysphagia. He denied pain or dysarthria and had not experienced recent neck trauma or had any recent operations. Ten years earlier, he developed infectious mononucleosis and remained healthy until a second episode of mononucleosis that began two days prior to his presenting symptoms. On examination, the tongue's left side appeared atrophic, without fasciculations and it deviated leftward upon protrusion. The cranial nerve examination was otherwise unremarkable. Notably, cerebellar testing was within normal limits and there were no other focal neurologic deficits.

Repeated MRIs over a 3-month period failed to demonstrate a structural lesion accounting for the patient's presentation. After 3 months, the patient improved clinically. His tongue mobility and swallowing returned to normal, although he subjectively felt left sided tongue thickening. Based on his clinical course and investigations, a diagnosis of post-infectious hypoglossal nerve palsy was made.

The hypoglossal nerve is a purely motor cranial nerve innervating the genioglossus, styloglossus and hyoglossus muscles in the tongue. Pathology affecting the pathway within

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and beyond the hypoglossal nucleus may present as ipsilateral tongue paresis, tongue deviation towards the side of the lesion, atrophy, dysphagia, or dysarthria(2). There are many potential causes of hypoglossal nerve palsy. The largest review, a retrospective case series of 100 cases, reported tumors, predominantly malignant, as the most common cause(1). The most common malignancies reported were metastases, chordomas, nasopharyngeal carcinomas and lymphoma. Benign space-occupying lesions causing this pathology include schwannomas, meningiomas, ependymomas and craniopharyngiomas(1). Trauma is the next most common etiology, such as gun shot wounds or blunt injury(1). Fractures of the occipital condyle and odontoid process can disrupt the hypoglossal canal. Vascular causes include internal carotid and vertebral artery dissections or ectasia, vascular insufficiency(1), and dural arteriovenous fistulas(3). Systemic processes such as Guillain Barre Syndrome, multiple sclerosis, and diabetes mellitus are rare causes(1). Certain infections can cause hypoglossal nerve palsy including meningitis, osteomyelitis(1), poliomyelitis, syphilis, herpes simplex virus, cytomegalovirus and Epstein-Barr virus(3). Given that idiopathic isolated hypoglossal nerve palsy is a diagnosis of exclusion, a thorough investigative work-up should aim to rule out all of the above entities(3).

Hypoglossal nerve palsy due to infectious mononucleosis is a rare etiology; only six cases have been reported previously(4,5). The typical clinical presentation is an isolated, unilateral hypoglossal nerve palsy that occurs in children and adolescents. To our knowledge, this is the first case reported in a middle-aged male.

The diagnosis of hypoglossal nerve palsy requires an understanding of the differential diagnosis to guide history-

taking, physical examination and investigations. Precipitating events such as trauma, recent surgery, or infection should be noted. Associated signs and symptoms can narrow the differential. Multiple cranial nerve deficits, incoordination or nystagmus may indicate a structural lesion or vascular cause. Systemic sequelae of inflammatory or metabolic conditions can raise suspicion of rheumatoid arthritis, ankylosing spondylitis or diabetes mellitus.

The investigative work-up should begin with a high resolution MRI as structural lesions are the most common cause. Magnetic resonance angiography can reveal vertebral or carotid artery dissection or ectasia. If inflammatory or metabolic causes are suspected, serologic testing for autoimmune disease or routine biochemistry can be ordered. A positive Monospot or elevated EBV titers can identify infectious mononucleosis. If after thorough investigation no clear etiology of the hypoglossal palsy can be found, then the possibility of isolated idiopathic hypoglossal nerve palsy remains.

Infectious mononucleosis is a rare cause of hypoglossal nerve palsy but should be suspected in isolated, unilateral clinical presentations. There is no accepted management of post-infectious hypoglossal nerve palsy due to its rarity. Of the six cases reported, corticosteroid therapy was initiated in three cases. Five patients resolved completely(4); only one case resulted in a persistent deficit after pulse steroids(5). Based on

the limited literature available, hypoglossal nerve injury post-mononucleosis infection appears to be a benign self-limiting condition. The few case reports available suggest that once the more serious potential etiologies have been excluded, no further treatment is required.

*Daniel Mendelsohn, Faizal Haji, Wai P. Ng*  
University of Western Ontario, London, Ontario, Canada

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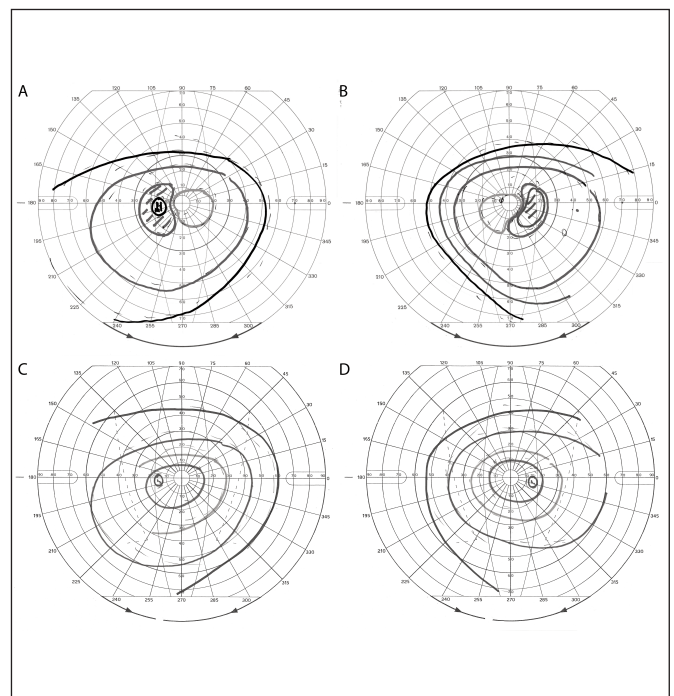
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#### TO THE EDITOR

#### Melanoma-Associated Retinopathy Report of a Case and Review

Melanoma associated retinopathy (MAR) is a rare visual autoimmune condition associated with metastatic malignant melanoma (MM). The case of a patient with spontaneous MAR resolution and a review of the literature are presented.

A 67-year-old female was diagnosed with metastatic subcutaneous MM from unknown primary in 1995. She was treated by surgical resection but developed pulmonary metastasis in May 2001. She underwent wedge resection of lung nodules and was enrolled in a phase I melanoma vaccine trial. Two weeks after surgery, she began experiencing bilateral white sparkling light sensations described as similar to "looking through a lace curtain" along with a decreased night vision preventing her from driving. Visual acuity was 20/25 on the right and 20/40 on the left. Neurological examination was otherwise unremarkable. Goldmann perimetry revealed bilateral constriction of the temporal fields (Figure 1, A-B). Brain MRI was only remarkable for an incidental left parietal meningioma and electroencephalogram recording was normal. ERG demonstrated decreased dark-adapted ERG b-wave amplitude consistent with MAR. After 9 years of continuous visual symptoms and no evidence of MM recurrence, she reported resolution her photopsia in August 2010. On March 2011, she had complete resolution of temporal fields' constriction (Figure 1, C-D) and stable visual acuity.



**Figure:** Goldmann Perimetry. A: Right eye; constriction of temporal field. B: Left eye; constriction of temporal field. C: Right eye; return to normal visual field. D: Left eye; return to normal visual field.