Dysarthria and dysphagia due to the opercular syndrome in multiple sclerosis

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Abstract

We report a patient with relapsing-remitting multiple sclerosis (RRMS), who developed bilateral severe tongue weakness due to the anterior opercular syndrome. This was caused by a recent inflammatory demyelinating lesion in the right perisylvian juxtacortical region, superimposed on a pre-existing left perisylvian lesion, which had previously caused temporary isolated right tongue weakness.

Keywords: dysarthria; dysphagia; Foix-Chavany-Marie syndrome; glossoplegia; multiple sclerosis; opercular syndrome

Introduction

The operculum is the cerebral cortex and subcortical white matter covering the insula. Bilateral anterior perisylvian lesions, involving the primary motor cortex within the frontal and parietal opercula, can produce a syndrome termed the anterior opercular syndrome (or Foix-Chavany-Marie syndrome) [1]. In its fully developed form, the syndrome is characterised by voluntary paralysis of the masticatory, facial, pharyngeal and lingual muscles, with preservation of emotional and automatic movements. In adults, the syndrome is usually caused by cerebral infarction [1], but may also be due to other disease processes, including herpes simplex encephalitis [2]. To our knowledge, the syndrome has not been reported in multiple sclerosis (MS). Here, we report a patient with MS who developed severe dysarthria and dysphagia due to glossoplegia caused by the anterior opercular syndrome.

Case report

A 34-year-old female with established relapsing-remitting (RR) MS presented, in March 2006, with slurred speech and impaired tongue movement, which had commenced seven days previously. She described slowing of tongue movement, causing food to occasionally lodge in her left cheek, and difficulty in moving a food bolus towards the back of her mouth. On examination, the patient had moderately severe dysarthria. Her tongue deviated to the left on protrusion, and movement on the left side of the tongue was of reduced range, rate and strength. Her symptoms and signs improved after 24 hours whilst in the air-conditioned hospital, and she was discharged.

Over the following two days, she gradually noted worsening of speech disturbance, and new symptoms of left facial droop and difficulty swallowing. The patient was only able to swallow sips of liquid, and her husband had noted that her speech had become almost unintelligible. On examination, she had severe dysarthria, a pathologically brisk jaw jerk, and complete left upper motor neuron voluntary facial weakness with preserved emotional facial expression. She had bilateral voluntary palatal weakness, worse on the left with the palate moving to the right. The gag reflex was preserved. There was virtual glossoplegia with severe weakness of the right tongue, and complete paralysis of the left tongue. Assessment by a speech pathologist revealed
dysarthria and oral-stage dysphagia with a normal pharyngeal phase.

A magnetic resonance imaging (MRI) brain scan at this time showed multiple cerebral white matter lesions hyperintense on FLAIR imaging, including periventricular and juxtacortical lesions. There was a large lesion involving the right perisylvian juxtacortical white matter (Figure 1A), which enhanced with gadolinium (Figure 1B). In addition, there was a lesion on FLAIR imaging in the left perisylvian region (Figure 1A). There were no lesions in the brainstem or cerebellum. The patient was treated with intravenous methylprednisolone therapy (1 g) daily for three days. Her speech and swallowing gradually improved.

The patient’s first symptoms of MS had commenced in 1999, when she developed a left hemiparesis. A MRI brain scan at that time revealed several cerebral white matter lesions, including a deep right cerebral lesion. Cerebrospinal fluid (CSF) examination revealed a leukocyte count of \(5 \times 10^6/L\), and oligoclonal immunoglobulin G bands in the CSF, but not the serum. Her symptoms improved spontaneously after six weeks. In February 2000, she experienced weakness of the left face, left upper limb and both lower limbs, lasting for one week. In March 2000, she had bilateral visual impairment for 10 days. A diagnosis of RRMS was made, and the patient was commenced on subcutaneous interferon beta-1b therapy. She noted an increase in depressive symptoms whilst on interferon beta-1b, and ceased this in June 2004. She was then commenced on subcutaneous glatiramer acetate.

In November 2004, the patient developed slurred speech and difficulty moving food around with her tongue due to isolated right tongue weakness. On that occasion, food accumulated in the right side of her mouth, and neurological examination showed weakness of the right side of her tongue. A MRI brain scan in March 2005 revealed the same lesion in the left perisylvian region as the scan in March 2006, but no lesion in the right perisylvian region. She was treated with intravenous methylprednisolone (500 mg) daily for five days and her symptoms improved.

![Figure 1](A) MRI brain scan axial FLAIR image showing a right juxtacortical perisylvian lesion (arrow), and a smaller left perisylvian lesion. (B) MRI brain scan coronal T1-weighted image showing gadolinium enhancement in the right perisylvian region (arrow).

**Discussion**

Our patient with RRMS developed bilateral severe tongue weakness causing severe dysarthria and oral-stage dysphagia. This was associated with bilateral upper motor
neuron palatal weakness, and left voluntary facial weakness with preserved emotional expression. These clinical features are explained by the anterior opercular syndrome due to a recent inflammatory demyelinating lesion in the right perisylvian juxtacortical region, superimposed upon a previous left perisylvian juxtacortical lesion.

In normal subjects, the right and left halves of the tongue each receive innervation from the primary motor cortex of each cerebral hemisphere, with the crossed corticolingual pathway activating a larger motor response that the uncrossed pathway [3]. Interruption of the cortical innervation from one hemisphere may produce only mild contralateral tongue weakness without dysarthria because of compensatory overactivation of the tongue muscles by the intact uncrossed corticolingual pathway from the normal hemisphere [3]. In the anterior opercular syndrome, bilateral anterior perisylvian lesions interrupt the cortical innervation from both hemispheres and result in severe bilateral tongue weakness and dysarthria, as in our patient. Similar mechanisms underpin the volitional weakness of the other cranial muscles in the anterior opercular syndrome. The preservation of emotional expression in this syndrome is explained by activation of the cranial muscles by pathways arising from outside the primary motor cortex [4].

Dysarthria and dysphagia are common symptoms of MS, and are usually attributed to brainstem or cerebellar involvement. Indeed, our patient’s previous episode of isolated right-sided tongue weakness in November 2004 was initially thought to be due to a brainstem lesion. In retrospect, it is clear that this was due to a lesion in the left perisylvian region.

To our knowledge, the anterior opercular syndrome has not previously been reported in MS, although it has been described in a child with acute disseminated encephalomyelitis [5], and there is a report of unilateral volitional facial and glossal paresis due to a large contralateral ‘frontocentral’ white matter lesion in MS [4]. Our case highlights the need to consider cerebral cortical and juxtacortical lesions, as well as brainstem and cerebellar lesions, as a cause for dysarthria and dysphagia in MS.

References