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Lingual epilepsia partialis continua in Rasmussen's encephalitis

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ABSTRACT – We report an adult male who presented with disabling dysarthria due to epilepsia partialis continua (EPC) of the left half of the tongue. The clinical, brain magnetic resonance imaging and electroencephalographic features were consistent with Rasmussen's encephalitis, although, despite having had the disease for over 15 years, he did not exhibit any hemiparesis. The accompanying video illustrates the lingual EPC and its total resolution following a right frontal opercular focal cortical resection.
[Published with video sequences].

Keywords: Rasmussen's encephalitis, epilepsia partialis continua, tongue, lingual EPC

Epilepsia partialis continua (EPC) has been defined as spontaneous, regular or irregular, focal clonic muscular twitches of cerebral cortical origin continuing for hours, days, weeks or months (Thomas *et al.* 1977). EPC can occur in acute as well as chronic lesions of the brain and of inflammatory, dysplastic, vascular, neoplastic, demyelinating, traumatic, and metabolic etiology (Thomas *et al.* 1977, Pandian *et al.* 2002). Chronic EPC continuing for months or years with onset in childhood or adolescent age group is characteristic of focal malformations of cortical development (Nakken *et al.* 2005) and Rasmussen's encephalitis (Bien *et al.* 2005).

EPC may afflict any muscle of the body; however, facial and distal limb muscles are preferentially involved (Thomas *et al.* 1977). Isolated EPC of the tongue (lingual EPC) is extremely uncommon (Thomas *et al.* 1977, Jabbari and Coker 1981, Thomas *et al.* 1995), and, to our knowledge, has not been reported in Rasmussen's encephalitis. This prompted us to document a patient with Rasmussen's encephalitis with lingual EPC which was treated successfully by focal resection of the cortical tongue area.

Case report

The patient is a 25-year-old, right-handed male who had an uneventful birth and early development, whose recurrent unprovoked left focal seizures began at the age of 9 years. A feeling of heaviness in the left upper and lower extremities followed by jerking of the left face, left upper and lower extremities lasting for 2 to 3 minutes occurred initially at a rate of 2 to 3 per day, but gradually became more frequent.

From the age of 15 years, the patient had nearly continuous twitching of the

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left side of the face and distal left upper extremities, despite treatment with varying combinations of antiepileptic drugs. At least twice monthly, he required hospitalization for frequent left motor seizures with secondary generalization. Other than transient mild weakness of the left upper limb following a prolonged bout of focal motor seizures, he had not developed any persisting focal neurological deficit.

From the age of 20, the seizures gradually became less frequent and more restricted in distribution. When first seen by us in July 2002, at the age of 22, the patient exhibited EPC involving the left half of the tongue and left facial muscles, which at times spread to distal muscles of the left upper limb. The soft palate was not involved. The lingual EPC disturbed his articulation considerably. There was no weakness or sensory deficit afflicting the left-sided extremities. The tendon reflexes were symmetric and the remainder of the neurological examination was normal. We reviewed serial brain magnetic resonance imaging (MRI) scans, which revealed progressive atrophy of the right perisylvian area and caudate nucleus, and hyperintensities involving the right lentiform and caudate nuclei on T2-weighted sequences (*figure 1*). Video-scalp electroencephalographic monitoring recorded right central, temporal and parietal spikes and sharp waves over a slowed and attenuated background activity, and partial motor seizures of right hemispheric origin. Based on the above clinical, radiological and electrographic features, we made the etiological diagnosis of probable Rasmussen's encephalitis. We optimized his antiepileptic medication to a combination of oxcarbazepine (1200 mg/d), topiramate (300 mg/d), and clobazam (15 mg/d). In view of the long duration of the disorder and its abating course, we did not consider immunotherapy with corticosteroids or immunoglobulin as a worthwhile treatment option.

During the subsequent 3 years under our observation, the patient's focal seizures reduced considerably and they no longer involved the face and left upper limb. However, the continuous twitching of the left half of tongue at about 1 Hz persisted and interfered with his speech. Repeat brain MRIs showed further progression of the right perisylvian hemispheric atrophy with insular hyperintensity (*figure 1*). Functional MRI lateralized the language function to left hemisphere. Since his most disabling symptom was the EPC of the left half of the tongue interfering with his articulation, in September 2005, we undertook a focal resection of the right cerebral cortex subserving the tongue function.

Surgical procedure

A right frontoparietal craniotomy was performed under local anesthesia, with the patient remaining awake. Intraoperative electrocorticography (ECoG) defined the maximal spiking zone over the right fronto-parietal cortex. Intraoperative cortical stimulation was carried out to define the motor areas. The cortical area representing the tongue in the right frontal operculum was then carefully resected under constant clinical monitoring. A complete cessation of the tongue EPC was achieved (see video sequence), with minimal paresis of the tongue and the left half of face. Post-resection ECoG showed persistence of spikes over the right posterior parietal region. Since the objective of the operation was to stop the lingual EPC, further resection was not carried out in order to avoid neurological deficits. Neuropathology revealed shrunken neurons with pyknotic nuclei and gliosis, but chronic inflammatory cell infiltration was not seen.

Post-operatively, his articulation improved considerably. There was a mild supranuclear left facial paresis and the jaw deviated slightly to the left on opening the mouth. However, he could move the tongue from side-to-side and it retained a mainly midline position on protrusion. He could chew and swallow normally. When seen recently at 3-month, post-operative follow-up, he had mild, left central facial paresis, normal tongue movements and normal speech. The lingual EPC has not recurred (see video sequence).

Discussion

The clinical, MRI and EEG features in our patient agreed with the recently proposed European Consensus Criteria of RE (Bien *et al.* 2005). However, the absence of hemiparesis, despite having had the disease for over 15 years, is quite atypical of RE. Furthermore, the histopathological findings in our patient did not reveal the microglial nodules and perivascular chronic inflammatory cell infiltration characteristic of RE. The natural history of RE, as we understand today, is based on a relatively small number of patients with this condition referred to tertiary epilepsy centers for presurgical evaluation, and therefore is biased in favor of more severely affected patients (Bien *et al.* 2002). The nonspecific nature of antibodies to glutamate receptor GluR3 (Mantegazza *et al.* 2002), which was initially proposed to be associated with RE (Rogers *et al.* 1994), and the recent identification of antibodies to α-7-acetylcholine receptor in patients with biopsy-proven RE (Watson *et al.* 2005) suggests that the syndrome of RE may encompass several different autoimmune entities and the clinical spectrum of RE may overlap with other autoimmune-mediated epilepsy syndromes. With the description of more and more patients with RE, the high variability in the clinical course and neurological manifestations are becoming increasingly apparent (Bien *et al.* 2002, Bien *et al.* 2005). Because of the multifocal distribution and marked heterogeneity of the pathological process (Pardo *et al.* 2004), a false negative result based on the examination of a limited surgical tissue, as was available in our patient, is not unexpected.

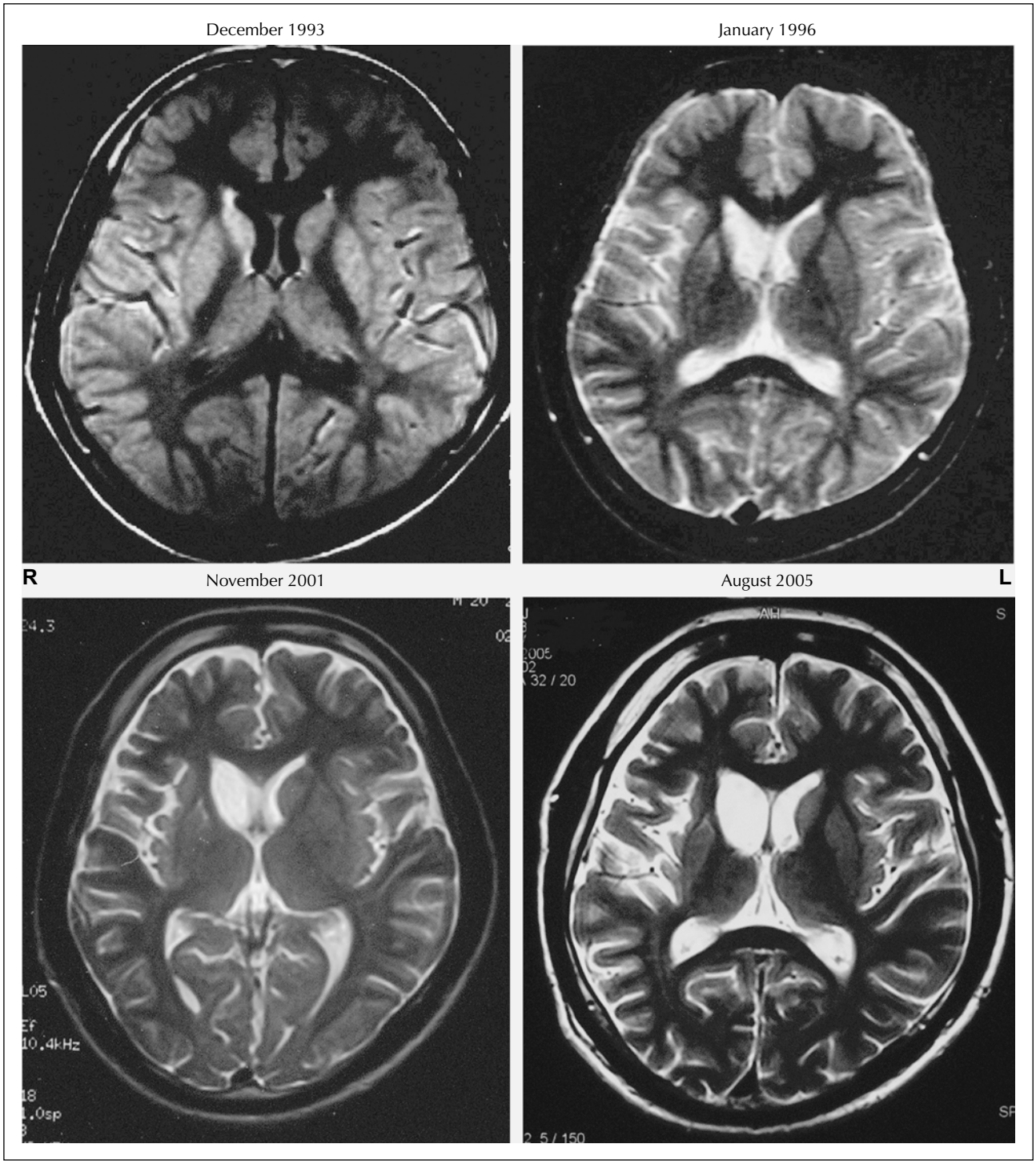

Figure 1. Serial brain MRI, FLAIR and T2-weighted sequences, reveal hyperintense signal in the right caduate nucleus initially, followed by progressive atrophy of right caudate and lentiform nuclei and dilation of the frontal horn of right lateral ventricle, and right hemispheric atrophy predominantly involving the perisylvian region.

Paroxysmal lingual myoclonus, often with involvement of the soft palate, jaw and facial muscles, as an epileptic phenomenon due to lesions involving the frontal opercular region, has been described (Thomas *et al.* 1977, Jabbari and Coker 1981, Thomas *et al.* 1995). The continuous lingual and palatal movements may interfere with articu-

lation and may even produce anarthria. Thomas *et al.* (1995) described three such patients having vascular and tumoral lesions involving the opercular region, under the title "opercular myoclonic-anarthric status epilepticus". Jabbari and Coker (1981) reported three children with rhythmic movements confined to the tongue, occurring mainly during sleep and accompanied by EEG desynchronization. The lingual EPC started in infancy and these children manifested features of diffuse brain disease. Synchronous movements of tongue, larynx and facial muscles may also occur as a nonepileptic phenomenon in palatal myoclonus syndrome due to focal brain stem lesions disrupting the dentatoolivary pathway (Lapresle 1986). Although EPC occurs in the majority of patients with RE at some time during the disease course (Bien *et al*, 2005), our patient with RE and isolated lingual EPC is unique.

EPC involving the facial and lingual musculature has been treated successfully by focal cortical resection (Lehman *et al.* 1994). In a series of 20 patients with seizure onset in the sensorimotor face area of the pre- and post-central gyri, resection of these areas resulted in significant reduction in seizures, with negligible neurological deficits. The results of focal resection in RE patients have been disappointing, and hemispherectomy in one of its modern forms is considered the only surgical option to achieve seizure-freedom in RE patients presenting with disabling hemiparesis (Bien *et al.* 2005). Our patient illustrates that, in those with no neurological deficit, medically refractory focal seizures in RE may be abolished by focal resection. We acknowledge that prolonged follow-up will be required to assess the impact of limited resection on sustained seizure control. □

References

Bien CG, Widman G, Urbach H, *et al.* The natural history of Rasmussen's encephalitis. *Brain* 2002; 125: 1751-9.

Bien CG, Granata T, Antozzi C, *et al.* Pathogenesis, diagnosis and treatment of Rasmussen encephalitis. A European consensus statement. *Brain* 2005; 128: 454-71.

Jabbari B, Coker SB. Paroxysmal, rhythmic lingual movements and chronic epilepsy. *Neurology* 1981; 31: 1364-7.

Lapresle J. Palatal myoclonus. *Adv Neurol* 1986; 43: 265-73.

Lehman R, Andermann F, Olivier A, *et al.* Seizures with onset in the sensorimotor face area: clinical patterns and results of surgical treatment in 20 patients. *Epilepsia* 1994; 35: 1117-24.

Mantegazza R, Bernasconi P, Baggi F, *et al.* Antibodies against GluR3 peptides are not specific for Rasmussen's encephalitis but are also present in epilepsy patients with severe, early onset disease and intractable seizures. *J Neuroimmunol* 2002; 131: 179-85.

Nakken KO, Server A, Kostov H, *et al.* A patient with a 44-year history of epilepsia partialis continua caused by a perirolandic cortical dysplasia. *Epilepsy Behav* 2005; 6: 94-7.

Pandian JD, Thomas SV, Santoshkumar B, *et al.* Epilepsia partialis continua: a clinical and electroencephalography study. *Seizure* 2002; 11: 437-41.

Pardo CA, Vining EPG, Guo L, *et al.* The pathology of Rasmussen syndrome: stages of cortical involvement and neuropathological studies in 45 hemispherectomies. *Epilepsia* 2004; 45: 516-26.

Rogers SW, Andrews PI, Gahring LC, *et al.* Autoantibodies to glutamate receptor GluR3 in Rasmussen's encephalitis. *Science* 1994; 265: 648-51.

Thomas JE, Reagan TJ, Klass DW. Epilepsia partialis continua. A review of 32 cases. *Arch Neurol* 1977; 34: 266-75.

Thomas P, Borg M, Suisse G, *et al.* Opercular myoclonic-anarthric status epilepticus. *Epilepsia* 1995; 36: 281-9.

Watson R, Jepson JEC, Bermudez I, *et al.* α-7-Acetylcholine receptor antibodies in two patients with Rasmussen's encephalitis. *Neurology* 2005; 65: 1802-4.

Legend for video sequences

Intraoperative recording during right frontal opercular focal cortical resection under awake craniotomy. Pre-resection recording shows rhythmic movements at 1 Hz involving the left half of the tongue. Immediately following resection, these movements were totally abolished, and the patient could protrude and move the tongue. At 3-month follow-up, mild left central facial weakness, slight deviation of the tongue to the left on protrusion, absence of EPC, normal side-to-side movement of the tongue and normal speech are evident.