

Two cases of opercular myoclonic-anarthric status epilepticus

Amaia Muñoz-Lopetegi, Gorka Fernández García de Eulate, Jon Rodríguez-Antigüedad Muñoz, Alberto Bergareche, Juan José Poza

Department of Neurology, Donostia University Hospital, San Sebastián, Spain

Received May 10, 2017; Accepted February 06, 2018

ABSTRACT – Opercular myoclonic-anarthric status epilepticus (OMASE) is a rare form of epilepsy partialis continua presenting as fluctuating dysarthria, or even anarthria. The condition is caused by an epileptogenic lesion involving the opercular cortex of either hemisphere. Speech impairment is secondary to bilateral epileptic activity affecting the glossopharyngeal muscles. This bilateral nature of the condition is due to the fact that innervation of cranial nerves V, VII, IX, X and XII from the opercular area of the primary motor cortex is bilateral. The aetiology of the condition varies, and includes vascular lesions, tumours, and encephalitis, among other causes. A low threshold for clinical suspicion is necessary in order to ensure the timely initiation of antiepileptic treatment, thereby preventing the condition from becoming drug resistant. We present two cases of OMASE which differ in terms of aetiology, clinical course, and treatment response. [*Published with video sequences on www.epilepticdisorders.com*].

Key words: epilepsy partialis continua, glossopharyngeal musculature, status epilepticus, dysarthria, anarthria

Opercular myoclonic-anarthric status epilepticus (OMASE) is characterised by fluctuating cortical dysarthria, and even anarthria, associated with epileptic myoclonus involving the glossopharyngeal muscles bilaterally. The condition is considered a form of epilepsy partialis continua in which epileptic activity can originate in either operculum. We present two cases of OMASE.

Case 1

A 78-year-old man attended the emergency department due to

difficulty in speaking. Symptom onset was sudden and followed by clonic movements on the left side of his lips. He had a history of high blood pressure and atrial fibrillation and was receiving acenocoumarol, nifedipine, enalapril, and omeprazole. The patient had visited the emergency department two days previously due to difficulty in speaking, however, this episode spontaneously resolved within 15 minutes and was interpreted as a transient ischaemic attack.

During the clinical examination, the patient had a normal level of consciousness and obeyed commands,



VIDEOS ONLINE

Correspondence:

Amaia Muñoz-Lopetegi
Department of Neurology,
Donostia University Hospital,
P. Dr. Beguiristain 109. 20014,
San Sebastián, Spain
<a.munozlopetegi@gmail.com>



Figure 1. EEG of Case 1 slowing can be observed in the right fronto-temporal region, with no epileptiform activity.

but was unable to produce sound. Bilateral clonic movements involving the tongue and soft palate resulted in anarthria and severe dysphagia to both solids and liquids. The patient's international normalised ratio was 1.4.

A brain CT scan revealed a small hyperdense area in the right parietal lobe, suggestive of a haemorrhagic lesion. A CT angiogram ruled out occlusion of the major cerebral vessels. The image also revealed an area of increased vascularisation, with prominent vessels in the right fronto-temporal region. This corresponded to an area of increased blood volume and normal transit time on perfusion sequences.

An EEG (*figure 1*) displayed asymmetric background activity, with slowing in the right fronto-temporal region. No epileptiform activity was detected, despite the fact that pharyngeal clonic movements could be observed throughout the study.

A fibrolaryngoscopy (*video sequence 1*) showed bilateral, rhythmic, symmetric movements in the soft palate, the base of the tongue, and in both arytenoid cartilages. No abnormalities were observed in the vocal folds.

A brain MRI scan displayed a lesion affecting the insula and adjacent areas of the frontal, anterior parietal, and temporal lobes. The lesion appeared hyperintense on T2-weighted images and showed restricted diffusion, with a gyriform pattern. Two small haemorrhagic lesions were observed on the right parietal lobe. Extensive vasogenic oedema surrounded the lesion, although there was no gadolinium uptake (*figure 2*). CT venography ruled out venous occlusions.

The patient was started on 500 mg levetiracetam/12 hours. Clonic movements gradually disappeared and the dysarthria and dysphagia resolved.

The lesion was initially interpreted as an area of subacute ischaemia since it appeared to correspond to

the territory of the M2 segment of the middle cerebral artery. However, some abnormal features were observed, such as the extensive oedema and increased vascularisation within the region. A CSF analysis and a complete blood count were performed in order to rule out other possible aetiologies.

The blood test returned positive results for anti-nuclear antibodies (titre: 1:640, with a nucleolar pattern). Results for anti-ENA, anti-DNA, and anti-cardiolipin antibodies were negative. Serology results were negative; no neuronal surface antibodies or intracellular antibodies were detected. The study revealed no further remarkable information.

The CSF obtained from the lumbar puncture was clear, with 5 leukocytes/ μ L, 56 mg/dL proteins, and normal glucose levels. There was no intrathecal immunoglobulin synthesis; the tests for anti-neuronal and onconeural antibodies and the microbiology and serological studies also returned negative results. CSF flow cytometry and cytology results were normal.

The patient was discharged and instructed to continue taking 500 mg levetiracetam/12 hours.

A follow-up MRI scan performed one month after discharge revealed considerable growth of the lesion. The scan revealed contrast uptake and a change in the surrounding oedema, which had become more extensive (*figure 2*). The perfusion MRI and spectroscopy study results were not suggestive of glial tumour or lymphoma. However, this evidence is not sufficient to rule out these diagnoses. Given the slow progression and the fact that the region initially involved corresponded to a vascular territory, we considered the possibility that the lesion could be secondary to small-vessel vasculitis. However, evidence in support of this hypothesis was insufficient. Unfortunately, the patient was lost to follow-up.

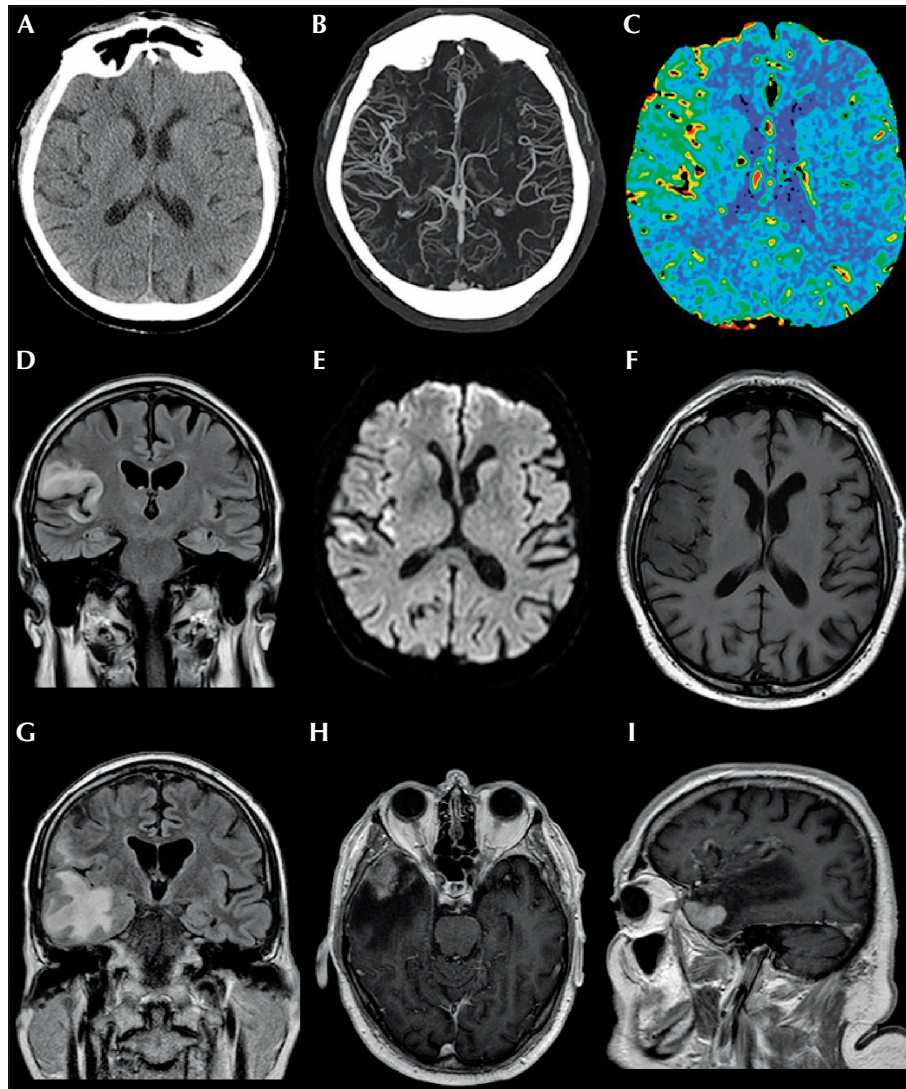


Figure 2. Neuroimaging studies of Case 1: CT and MR images taken at admission and follow-up MR images taken a month later. A) Non-enhanced brain CT performed at admission. B) CT angiogram taken at admission. C) CT perfusion sequence taken at admission. D) MRI coronal FLAIR sequence taken at admission. E) Diffusion-weighted axial MR image taken at admission. F) Gadolinium-enhanced axial T1-weighted sequence taken at admission. G) Follow-up coronal FLAIR sequence. H) Follow-up gadolinium-enhanced axial T1-weighted sequence. I) Follow-up gadolinium-enhanced sagittal T1-weighted sequence.

Case 2

A 75-year-old man attended the emergency department due to persistent left hemifacial clonic movements preventing correct speech. No other areas of the body were affected and focal neurological deficits were not observed during the examination. The patient's condition was interpreted as simple partial status epilepticus. He received 1,000 mg levetiracetam intravenously at the emergency department, and the clonic movements resolved. The patient was admitted to hospital.

He had had a generalised tonic seizure three years previously, and had since been receiving 500 mg levetiracetam/12 hours. A brain CT scan performed at the time of that seizure revealed no abnormalities. Two years later, the patient had sustained head trauma with right fronto-temporo-parietal subarachnoid haemorrhage, right fronto-temporal subdural haematoma, and haemorrhagic contusions in the anterior temporal, fronto-basal, and posterior fronto-temporal regions; the patient was treated surgically. The left hemifacial clonic movements reappeared several hours after admission, extending to the left arm.



Figure 3. EEG of Case 2 showing continuous epileptiform activity with pseudo-periodic spikes and sharp waves in the right temporal region, with slightly slowed background activity.

The patient initially responded to intravenous levetiracetam and clonazepam, but a different pattern of involuntary movements appeared several minutes later. Myoclonus involved the chin and tongue, which protruded continually and arrhythmically from the mouth, accompanied by drooling. The patient presented severe dysarthria and anarthria. His level of consciousness was normal, and he was able to understand and follow orders. The examination revealed no further abnormalities.

It was not possible to obtain an EEG reading until the third day after admission. The results showed epileptiform activity with pseudo-periodic spikes and sharp waves in the right temporal region, and slightly slowed background activity (*figure 3*).

This EEG activity manifested clinically as continuous movement of the chin (*video sequence 2*). Further EEG readings were taken on the fifth, ninth, and twelfth days after admission. Despite a degree of progressive improvement, EEG readings continued to show epileptiform activity.

A brain MRI scan displayed the known sequelae of the head trauma. Restricted diffusion, involving the right operculum, was also observed; this is suggestive of postictal changes (*figure 4*).

At admission, treatment was initiated with 1,000 mg levetiracetam/12 hours. The dose was increased to 1,500 mg/12 hours and lacosamide was added at a maintenance dose of 150 mg/12 hours. As the continuous myoclonic movements persisted, valproic acid was also added at a dose of 2,000 mg/day. Myoclonus progressively improved and disappeared on the seventh day after admission. However, EEG epileptiform activity persisted, and practical anarthria and severe

dysphagia remained until post-admission Day 13, when oral nutrition was finally possible.

The patient was diagnosed with OMASE secondary to sequelae of a traumatic brain injury.

Discussion

OMASE is a rare condition, with only five cases registered on the PubMed database: three cases were of vascular origin (Thomas *et al.*, 1995; Bhaskara *et al.*, 2013), one was secondary to autoimmune encephalitis (Zanotelli and Rodrigues, 2013), and one was secondary to a tumour (Thomas *et al.*, 1995). It is a form of simple partial motor status epilepticus that predominantly affects the oropharyngeal musculature. Unlike the more frequently described palatal myoclonus, which generally occurs when brainstem lesions interfere in the dentato-rubro-olivary pathway, OMASE is cortical in origin and responds to AEDs.

OMASE occurs when the opercular region of the inferior Rolandic cortex in either hemisphere is affected by an epileptogenic lesion, which may be of varying aetiology. The anterior operculum contains a large representation of the face in the motor homunculus. The bilateral, symmetric involvement of the facial and oropharyngeal musculature is explained by the bilateral cortical representation of cranial nerves V, VII, IX, X and XII. The presence of exclusively unilateral labial clonic movements is unsurprising, however, as this part of the facial musculature is represented only in the contralateral primary motor cortex.

Anarthria is an epileptic symptom caused by involuntary, myoclonic movements of the muscles involved in

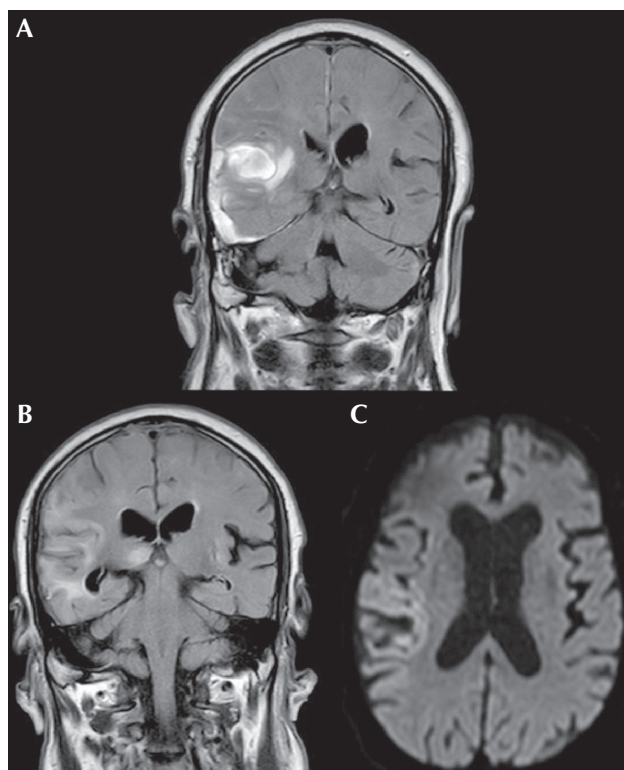


Figure 4. MRI of Case 2. A) FLAIR sequence performed two years previously, showing haemorrhagic contusions in the fronto-temporal region. B) FLAIR sequence for the episode reported, showing sequelae of the previous episode. C) Diffusion-weighted sequence showing cortical restriction, suggestive of postictal changes.

speech. However, it may also occur post-seizure in the context of facio-pharyngo-glosso-masticatory paresis, compatible with Todd paralysis. It may be considered a transient form of classic Foix-Chavany-Marie syndrome, also known as anterior opercular syndrome.

Pure anarthria also occurs in epileptic seizures originating in the primary negative motor area (adjacent to the primary motor area) or the supplementary motor area, also known as the secondary motor area. In these cases, the cause of anarthria would be a negative process, and we would not observe the myoclonus shown in the video. Neither would we expect to see the clonic movements of the contralateral hand and face, or dysphagia.

Even if there is clinical suspicion that the epileptic focus is of cortical origin, EEG does not always show epileptiform activity, as we report in Case 1. This may be because the operculum's deep location in the cortex prevents signal detection by surface electrodes.

An EMG would have helped to better characterise muscle activity, but unfortunately, this was not performed in either case.

Early detection of this epileptic phenomenon is important in order to ensure prompt initiation of

antiepileptic treatment and prevent the condition from becoming drug resistant. Treatment is the same as that administered for any other type of symptomatic focal epilepsy.

As mentioned previously, the aetiology of the condition varies. In Case 1, diagnosis of a tumour or vasculitic process was proposed, however, this could not be confirmed since the patient was lost to follow-up. In Case 2, the epileptic activity was secondary to a chronic traumatic lesion. □

Legends for video sequences

Video sequence 1

Case 1: fibrolaryngoscopy showing bilateral rhythmic, symmetric movements of the soft palate, base of the tongue, and both arytenoid cartilages. The patient was anarthric and had severe dysphagia.

Video sequence 2

Case 2: EEG recording during drooling and continuous involuntary movement of the chin. The patient had a normal level of consciousness and followed orders.

Key words for video research on www.epilepticdisorders.com

Phenomenology: status epilepticus (non-convulsive), motor seizure (simple), focal seizure not otherwise specified

Localisation: operculum (right), operculum (left) proposed

Syndrome: epilepsia partialis continua

Aetiology: unknown (video 1), head trauma (video 2)

Supplementary data.

Summary didactic slides are available on the www.epilepticdisorders.com website.

Acknowledgements and disclosures.

The authors acknowledge Esteve Farma for writing support (English translation of the article).

None of the authors have any conflict of interest to declare.

References

Bhaskara Rao JV, Vengamma B, Naveen T. Opercular myoclonic-anarthric status epilepticus: a report of two cases. *Ann Indian Acad Neurol* 2013; 16: 565-8.

Thomas P, Borg M, Suisse G, Chatel M. Opercularmyoclonic-anarthric status epilepticus. *Epilepsia* 1995; 36: 281-9.

Zanotelli B, Rodrigues T. Opercular myoclonic-anarthric status epilepticus due to glutamic acid decarboxylase antibody-associated encephalitis. *Epileptic Disord* 2013; 15: 342-6.

TEST YOURSELF



- (1) Motor activity, which affects glossopharyngeal muscles, is bilateral in OMASE. Can you describe the neuroanatomical substrate of this phenomenon?
- (2) Which two pathophysiological mechanisms can contribute to dysarthria or anarthria in this clinical picture?
- (3) Are there any lateralizing signs?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".