Clinical commentary

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Focal visual status epilepticus*

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ABSTRACT – Epileptic visual auras are elementary to complex and sometimes occur as colourful visual phenomena located close to or within the central part of the contralateral hemi-field. They typically last from seconds to a few minutes, which discriminates them from the usually longer-lasting visual auras (5-30 minutes) of patients suffering from migraine. We present an adult patient with occipital lobe epilepsy whose visual aura under epilepsy monitoring lasted for more than 30 minutes with almost no propagation, demonstrating a rare, but remarkable, sustained local epileptic network activity associated with resection of an occipital arterio-venous malformation.

Key words: epilepsy, status epilepticus, visual aura, occipital epilepsy, migraine, arterio-venous malformation

The majority of epileptic seizures last from seconds to a few minutes. A longer duration may point to an alternative cause such as psychogenic non-epileptic seizures or indicate status epilepticus, operationally defined as the persistence of a (tonic-clonic) seizure for more than five minutes or the occurrence of more than one (tonic-clonic) seizure without restoration of consciousness in between (Trinka *et al*., 2015). However, such pathophysiology-based conventions have not been fully established for non-convulsive status epilepticus without alteration of consciousness (Trinka *et al*., 2015). In addition, "epilepsia partialis continua" refers to a subgroup of focal status epilepticus with motor or non-motor phenomena, the latter alternatively termed "aura continua", lasting for at least 60 minutes (Mameniskiene *et al*., 2011).

Case study

A right-handed, 41-year-old woman had suffered an atypical intracerebral haemorrhage from an arterio-venous malformation (AVM) in the right occipital lobe two years previously. She reported a left paracentral visual scotoma but no other sequelae. Half a year after the event and after resection of the AVM, she experienced a visual aura that evolved into a tonic-clonic seizure.

Despite antiepileptic treatment, she reported experiencing visual auras on a weekly basis. She described them as bright, at times rotating, otherwise largely immobile,

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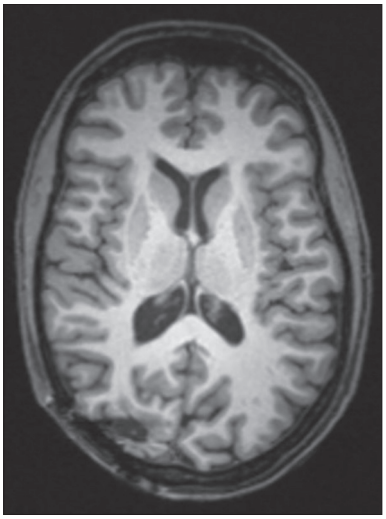

Figure 1. T1-weighed MRI demonstrating a right occipital scar two years after resection of an arterio-venous malformation.

sometimes colourful spots in the lower left visual field. These would often last for minutes, and she had noticed a duration of 15 minutes. She then occasionally developed epigastric and/or unpleasant olfactory sensations that could last for more than a minute. Every two months, on average, she suffered a tonic-clonic seizure. Several days after withdrawing her medication under epilepsy monitoring during pre-surgical workup, such a visual aura, lasting for 40 minutes and arising within her left lower visual field close to the permanent scotoma, occurred. The EEG seizure pattern remained highly localized to electrode O_2 and thus close to the right occipital scar throughout most of the seizure (*figures 1, 2*). After it spread to electrodes T6 and P4, clonazepam (1 mg; IV) was administered which quickly terminated the seizure. Source localization (solely based on a routine electrode montage with 20 electrodes) placed the epileptogenic source right of the pole of the right calcarine cortex, as expected by the highly localized seizure pattern. fMRI-retinotopy as well as tractography demonstrated a close relationship between epileptic scar and visual tract, indicating that a visual field defect may be regarded as an inevitable risk of potential occipital lobe epilepsy surgery in our case.

Discussion

Due to their duration, visual status epilepticus and visual epilepsia partialis continua may be difficult to differentiate from similar phenomena arising with migraine. In the clinical context, however, a definitive diagnosis is usually possible (Panayiotopoulos, 1999a; Eriksen *et al*., 2005; Hartl *et al*., 2017). Indeed, there are only very few reports of focal visual status epilepticus with symptoms lasting for longer than five minutes. These are summarized in *table 1*. Epileptic amaurosis which is poorly distinguished from postictal deficits and status epilepticus originating in the occipital lobe, where visual hallucinations are only the initial and not the predominating symptom, were excluded. In such cases, the criteria reported to be most reliable in differentiating between an epileptic and a migraine aura are the unilaterality of the former and the longer duration of the latter (Panayiotopoulos, 1999a). Eriksen *et al*. (2005) proposed a five-item score that assesses duration, symptom-dynamic, scotoma, fortification, and unilaterality in order to recognize visual auras associated with migraine and aid in clinical decision-making. Panayiotopoulos (1999b) found a prevalence of occipital lobe epilepsy of about 5%; 63 of his 1,360 epilepsy patients had occipital lobe epilepsy. The underlying conditions were early onset, benign childhood epilepsy (40%), idiopathic occipital epilepsy without photosensitivity, symptomatic occipital lobe epilepsy (25% each), and idiopathic photosensitive epilepsy (10%). Importantly, the prevalence of interictal epileptiform activity is particularly low. Based on neurosurgical series, only 20% of patients with symptomatic occipital lobe epilepsy exhibit interictal occipital epileptiform activity (Adcock and Panayiotopoulos, 2012). The same authors stated that "a well-localized unifocal rhythmic ictal discharge during occipital seizures is infrequent", indicating the rather unusual presentation in the current report (Adcock and Panayiotopoulos, 2012).

A search of the literature for similar cases yielded a report nearly identical to ours; a 42-year-old, right-handed woman with a history of epilepsy after embolization of a right parieto-occipital arterio-venous malformation, who had prolonged visual auras, one lasting for 13 minutes, documented during video-EEG monitoring. The lesion was eventually resected, and the patient remained seizure-free, albeit with impaired consciousness and occasional visual seizures (Hartl *et al*., 2015). Another similar case was recently published with an arterio-venous malformation again as the epileptogenic lesion. After embolization and complete resection of the lesion, the patient had been seizure-free for three years without any further neurological deficits (Strzelczyk *et al*., 2017). This may be of relevance to the issue of the long-lasting focality of the ictal discharges. While in our case, withdrawal of the antiepileptic medication certainly did increase the chance of occurrence and a longer duration of seizures, it did not induce rapid propagation as often seen after withdrawal of anticonvulsive therapy. The mechanisms by which epileptic activity is maintained within a focal network are not well defined. Apparently, epileptogenic tissue in these cases is, or becomes,

Table 1. Case reports of EEG-confirmed visual status epilepticus; association with vascular malformation is highlighted in bold.

Maximum duration	Age of onset	Seizure characteristics	Imaging	EEG	Reference
1 minute every 10 minutes	83	Multicoloured spots in the lower right quadrant spreading over the whole right visual field	Left mesio-occipital possible **vascular malformation**	Left occipital seizure pattern	Spatt and Mamoli, 2000
13 minutes (V-EEG)	39	Pulsating spots over the entire visual field with predominance on the left, moving to the left and becoming greenish	Post resection of an **arterio-venous malformation,** right parieto-occipital	Right parieto-occipital seizure pattern (V-EEG)	Hartl *et al.*, 2015
17 minutes (V-EEG)	15	Flickering in the right visual field, followed by a contralateral scotoma and colourful visual sensations	Left occipital gliosis	Left occipital seizure pattern (V-EEG)	Hartl *et al.*, 2015
20-30 minutes	14	Transparent, flickering line in the middle of the right hemi-field, expanding over the whole right hemifield and becoming colourful	**Arterio-venous malformation,** left temporo-occipital	Left occipital seizure pattern (V-EEG)	Strzelczyk *et al.*, 2017
45 minutes	32	Hallucination of "red flashing lights" in the left hemifield	Right mesio-occipital encephalomalacia in cCT	Frequent biposterior spikes + spike-waves, dominant posterior temporal	Aldrich *et al.*, 1989
>1 h	-	Continuous whitish photomes at the outer rim of the visual field, sometimes evolving over the entire visual field	**Operated haemangioma**	Unrevealing EEG	Mameniskiene *et al.*, 2011

Table 1. Case reports of EEG-confirmed visual status epilepticus; association with vascular malformation is highlighted in bold (*Continued*).

Maximum duration	Age of onset	Seizure characteristics	Imaging	EEG	Reference
>1 h	-	Flickering in the left visual field	-	Right occipital epileptiform EEG activity	Mameniskiene *et al.*, 2011
>1 h	-	Flickering in the left visual field, repetitive 1-2-h episodes	Non-specified occipital lesion	Non-specific focal ictal EEG abnormalities	Mameniskiene *et al.*, 2011
>1 h	-	Visual hallucinations of lines and circles	-	Left occipital runs of spikes	Mameniskiene *et al.*, 2011
60-90 minutes	42	Yellow, red and blue spots	Right occipital lesion after mycoplasmic meningoencephalitis	Right occipital interictal sharp waves; right occipital seizure pattern	Jobst *et al.*, 2009
A few hours	60	Palinopsia or abnormally recurring visual imagery, macropsia, unformed hallucinations, hemianopia	Left temporooccipital **cavernous haemangioma**	Left occipital seizure pattern	Kawai *et al.*, 2006
Up to 2 days	3	Flickering lights, obfuscation of vision by red and green lights; or shimmering ellipsoid, silver lights ("like a camera flash")	Hemispheric asymmetry	Right occipital lobe seizure pattern; abnormal photic stimulation response	Walker *et al.*, 1995
>3 years	10	Visual learning disorder; no visual hallucination	MRI normal; but FDG-PET with prominent left occipital hypermetabolism	Right occipital status epilepticus	Sheth and Riggs, 1999

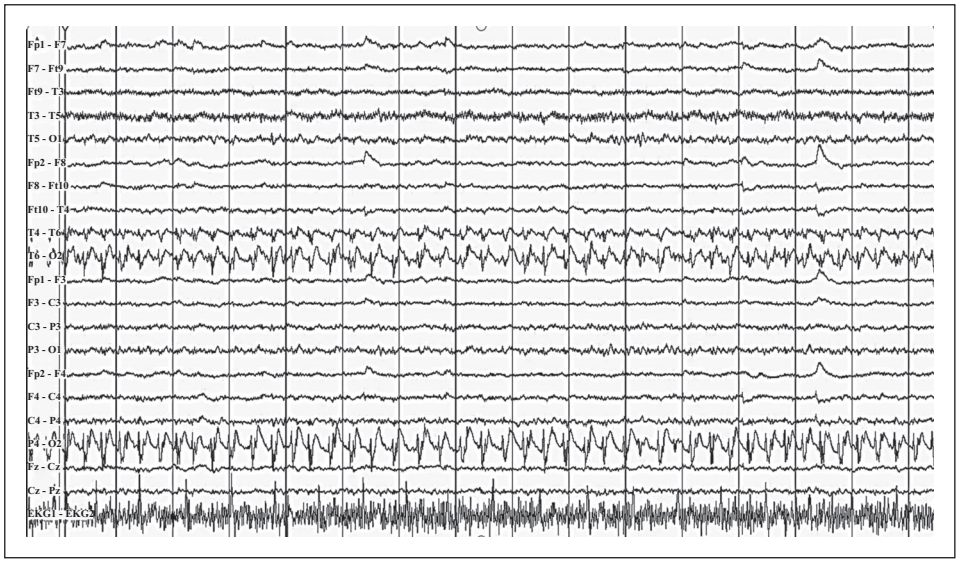

Figure 2. Well-localized EEG seizure pattern at electrode O_2.

electrically isolated. Isolation in such cases may be induced by proliferation of non-neurogenic tissue of inflammatory, glial or vascular origin. The high prevalence of vascular malformations in the cases presented in *table 1*, as well as in cases of amaurotic status epilepticus (Barry *et al*., 1985), indicate that such tissue, or scar tissue after its resection, may provide suitable conditions for focal status epilepticus. On the other hand, and taking into account the long duration of visual auras in migraine, conditions in the striate cortex itself may render spreading of electric activity less likely than in other cortices. Whether this property could be related to the macroscopic "stria" of Gennari -the dense stripe of highly myelinated horizontal fibres in layer IV of the primary visual cortex- remains to be shown.

Long-lasting epileptic visual auras illustrate the wide spectrum of propagation dynamics within epileptogenic networks and can be associated with vascular malformations. □

Disclosures.

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

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TEST YOURSELF

(1) Which clinical criteria differentiate best between a migrainous and an epileptic visual aura?

(2) Which clinical criteria currently define convulsive status epilepticus?

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".