Surgery of posterior fossa arachnoid cyst causing hydrocephalus unmasking the cause of epilepsy

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Abstract

Posterior fossa arachnoid cysts are benign collections of CSF within layers of arachnoid space and have a congenital origin. They are very rare and most often constitute incidental findings in imaging studies for several non-specific symptoms mainly headache and epilepsy. Epilepsy associated with arachnoid cysts in posterior fossa remains a controversial issue. We report a case of a symptomatic arachnoid cyst of posterior fossa, causing hydrocephalus in a patient with partial epilepsy, the structural cause of which was unmasked during epilepsy pre-surgical evaluation, and sufficient time elapsed for the shrinkage of the ventricles. This patient was admitted and treated at the University Hospital Center "Mother Teresa" in Tirana, Albania. This was an interesting case because the patient suffering from chronic epilepsy, presented with signs and symptoms of progressive intracranial hypertension caused by an arachnoid cyst in the posterior fossa that was managed successfully with surgical removal of the cyst wall. The possible cause of the patient's epilepsy was only hypothesized clinically, but no apparent structural cause was possible in preoperative imaging studies. We suggest that in posterior fossa arachnoid cysts or in posterior fossa cystic lesions causing hydrocephalus and associated with partial epilepsy, a supratentorial cause should always be sought before linking epilepsy with a possible, albeit rare cerebellar focus.

Key words: arachnoid cysts, posterior fossa arachnoid cysts, temporal lobe epilepsy.

Introduction

Arachnoid cysts are rare benign lesions representing 1% of all intracranial masses (1). They are collection of cerebrospinal fluid within layers of arachnoid space that do not communicate with the ventricular system (1). They are considered to be congenital in origin. The majority of these cysts are located in middle cranial fossa with a predilection of sylvian fissure (50%-60%), and most of these are only found incidentally on CT/MRI imaging (2). Headache and seizure are considered to be the most common symptoms. Posterior location of arachnoid cysts is rare and there is always concern to differentiate these cysts from other development abnormalities such as Dandy Walker Complex, Blake's Pouch cysts and Mega cisterna magna (3). These latter syndromes are frequently associated with disorders of other areas of the central nervous system such as malformations of cortical development and epilepsy. Epilepsy in the context of posterior arachnoid cysts has been a subject of case reports and it has been described in only four patients of a series of 2288 epilepsy patients (4). Moreover, the role of the cerebellum in epilepsy generation or participation in an epileptogenic network remains controversial. Arroyo et al. suggested that arachnoid cysts in patients with epilepsy do not necessarily reflect the location of the seizure focus (5). Whereas, Gan et al. using intraoperative electrocorticography (ECoG) demonstrated epileptic activity from the cerebellar tissue adjacent to an arachnoid cyst in one patient that improved after cyst fenestration, suggesting a possible relationship between epilepsy and posterior fossa lesions (6). There was evidence of focal epilepsy in posterior fossa lesions according to a case description, rather than a primary generalized form (4).

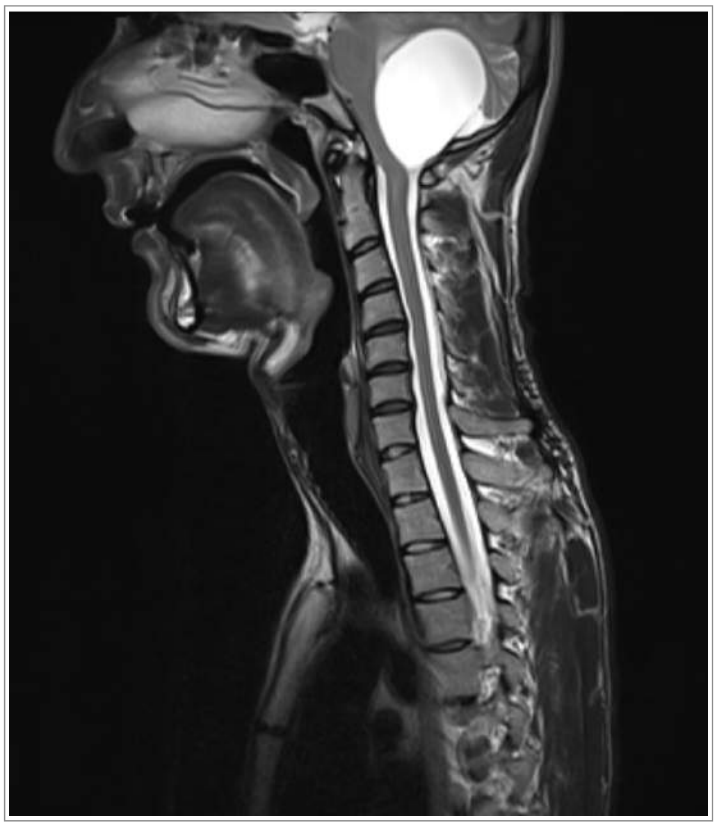
Case description

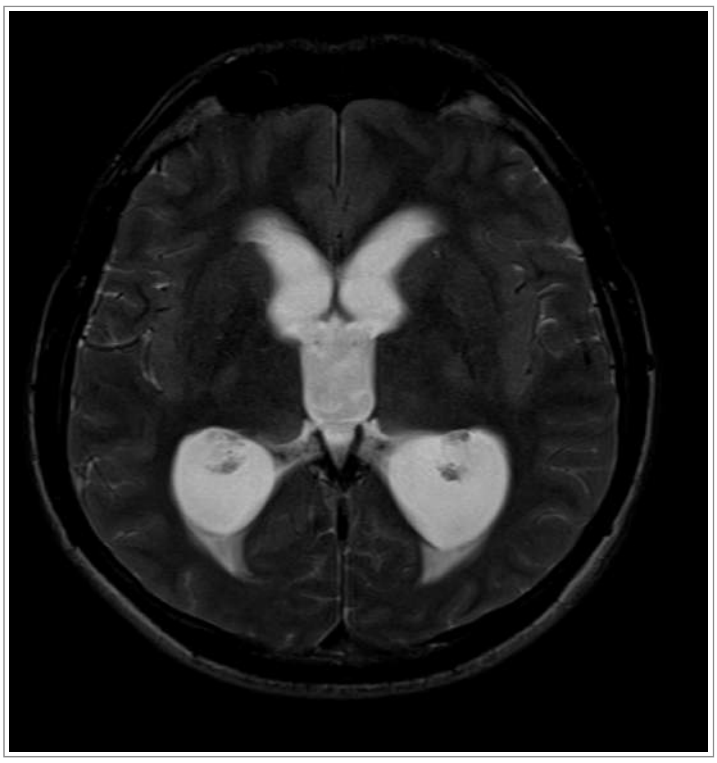
The patient was a 27-year old male, admitted at the University Hospital Center "Mother Teresa" in Tirana, Albania, with headache, vomiting, papilledema, torticollis (antecollis). Three months before admission intermittent headaches started to increase in frequency and severity, progressing to a syndrome of intracranial hypertension with headaches mainly in recumbent position, nausea, vomiting, neck stiffness and visual disturbances. His medical history was positive for an epilepsy disorder, starting seven years ago and drug resistant to carbamazepine 800 mg/day. Seizures were complex partial, starting with epigastric sensation, déjà vu phenomena and progressing to alteration of consciousness with oroalimentary and manual automatism associated with distonic posturing of the left arm with rare secondary generalizations. At the age of three years, he had experienced a possible febrile convulsion following high fever, but did well afterwards and developed normally.

Upon examination, the patient was distressed, maintained a flexed posture of the neck, and had papillary edema and no focal neurological deficits. MRI of the brain revealed an active triventricular hydrocephalus and a large cyst in the posterior cranial fossa. The cyst was symmetrically located in the midline posterior fossa. There was no contrast enhancement and the internal signal was isointense with cerebrospinal fluid intensity in T1, T2, FLAIR and no signal restriction in DWI images. The fluid was contained within a thin membrane visibile in the posterior and superior aspect of the cyst and had no communication with the fourth ventricle. It created an upward displacement of cerebellum and anterior displacement of medulla oblongata that appeared deformed and compressed anteriorly toward the clivus. The cyst descended to the foramen magnum. Imaging studies suggested an arachnoidal cyst. Supratentorial abnormalities were not present.

The patient was operated the following day of admission. A suboccipital craniotomy was performed in a semi-sitting position. Posterior wall of the cyst was removed with cysto-cisternostomy and specimens were sent for a pathological evaluation. The biopsy confirmed the macroscopic and imaging appearance of an arachnoidal cyst. The cyst wall

Figure 1. a) and b) Preoperative images demonstrating an arachnoidal cyst in the posterior cranial fossa producing enlargement of the ventricles

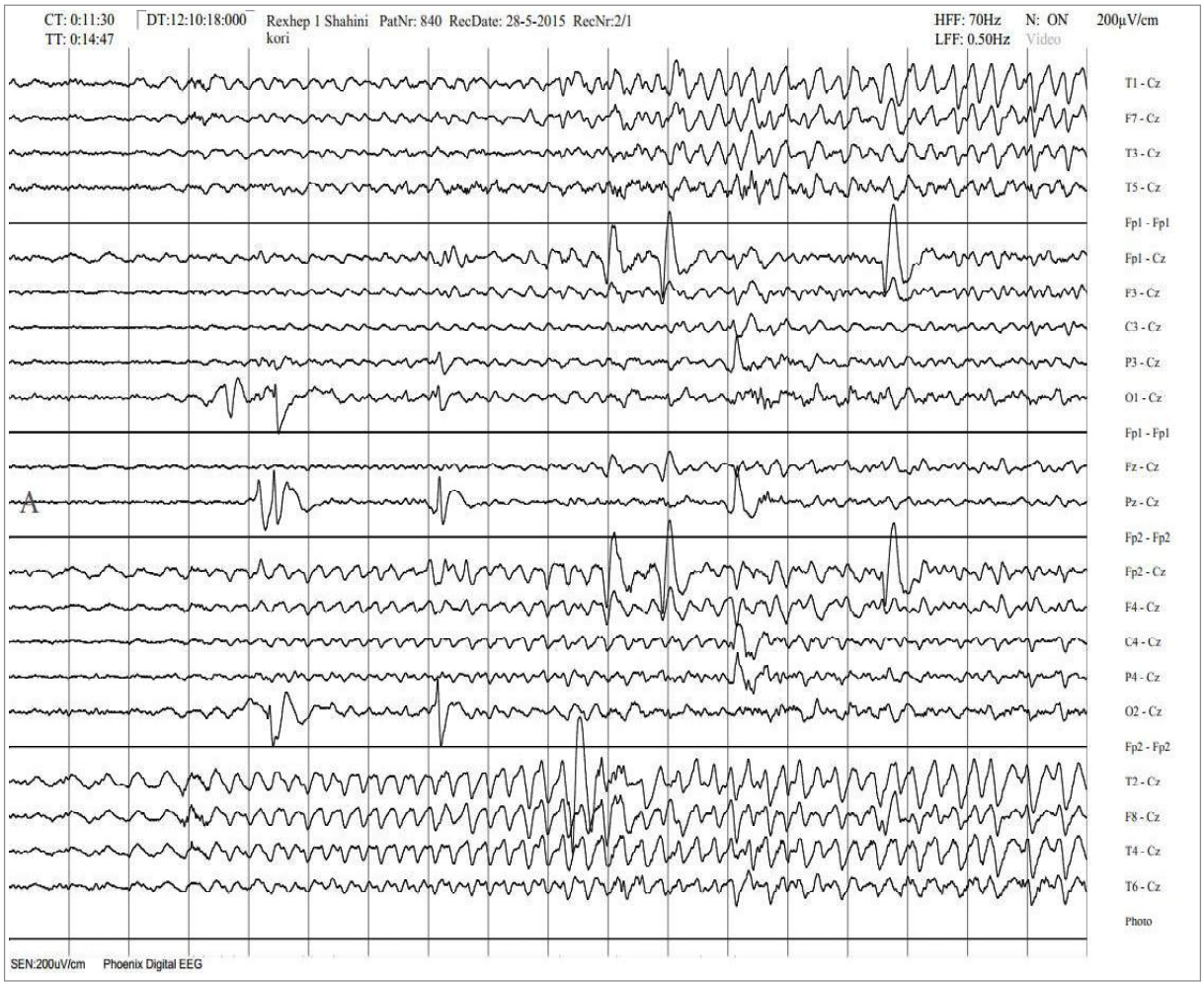


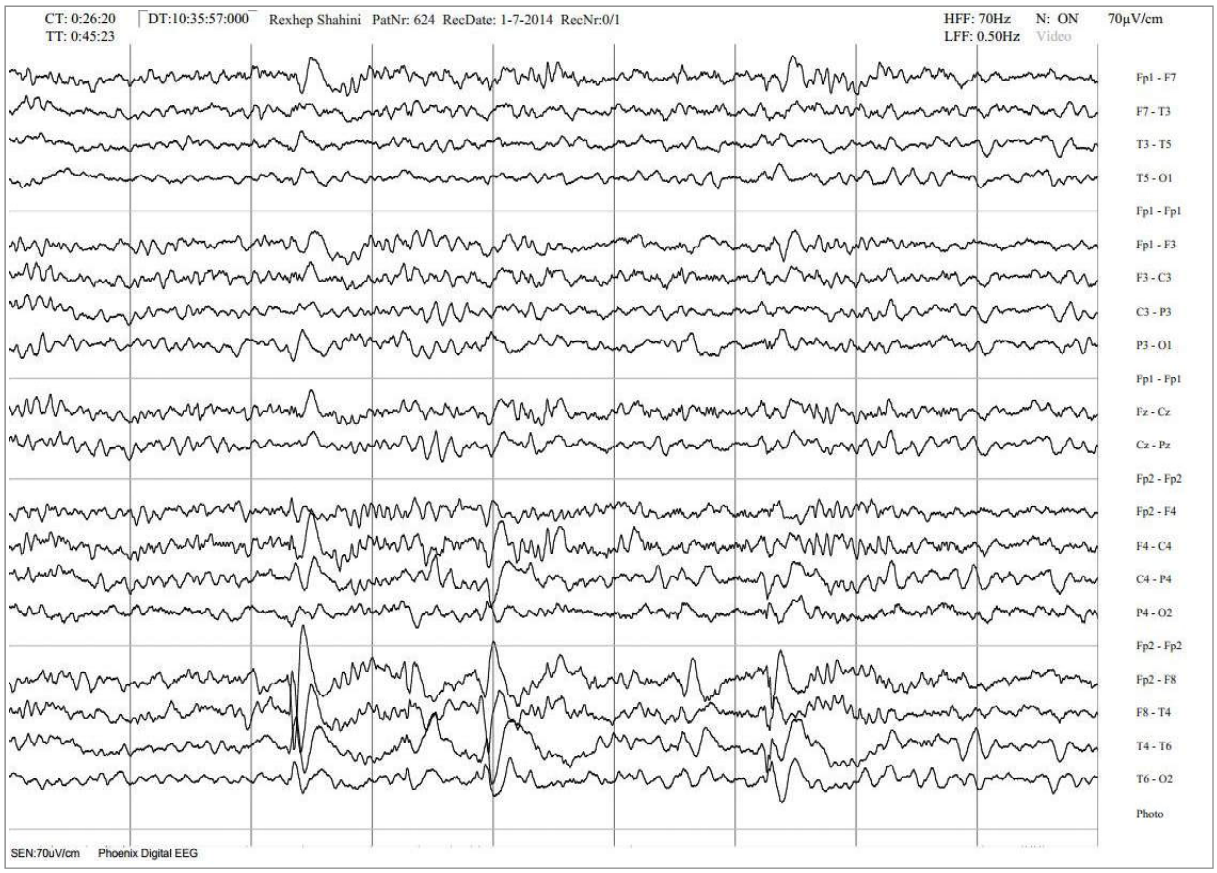


was lined with arachnoidal cells with an outer layer of connective tissue and no presence of neoplasia. The patient had immediate resolution of intracranial hypertension symptoms following surgery. After a normal postoperative CT scan the patient was discharged from the hospital the next day.

Because the patient still had seizures we proposed an evaluation with EEG that revealed focal epileptiform abnormalities in the right temporal lobe and less often abnormalities on the left temporal lobe. A second trial with another antiepileptic drug, valproate 1500 mg was proposed and the patient was seizure free only temporarily with seizure recrudescence after six months, hence becoming pharmacoresistant to two appropriately chosen AED. A pre-surgical evaluation for epilepsy surgery was proposed and ictal video-electroencephalography revealed a right temporal focus suggesting the origin of the seizures in the right medial temporal structures.

Figure 2. a) Interictal EEG demonstrating spike wave complexes in the right temporal anterior-temporo-basal region. b) Ictal video-EEG demonstrating an evolution of slow focal activity from delta to theta range starting in the right temporobasal (T2) electrode preceding clinical seizure by ten seconds (eye blink artefacts)

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Rexhep Shahini PatNr: 624 RecDate: 1-7-2014 RecNr:0/1
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LFF: 0.50Hz
N: ON
Video
70µV/cm
Fp1 - F7
F7 - T3
T3 - T5
T5 - O1
Fp1 - Fp1
Fp1 - F3
F3 - C3
C3 - P3
P3 - O1
Fp1 - Fp1
Fz - Cz
Cz - Pz
Fp2 - Fp2
Fp2 - F4
F4 - C4
C4 - P4
P4 - O2
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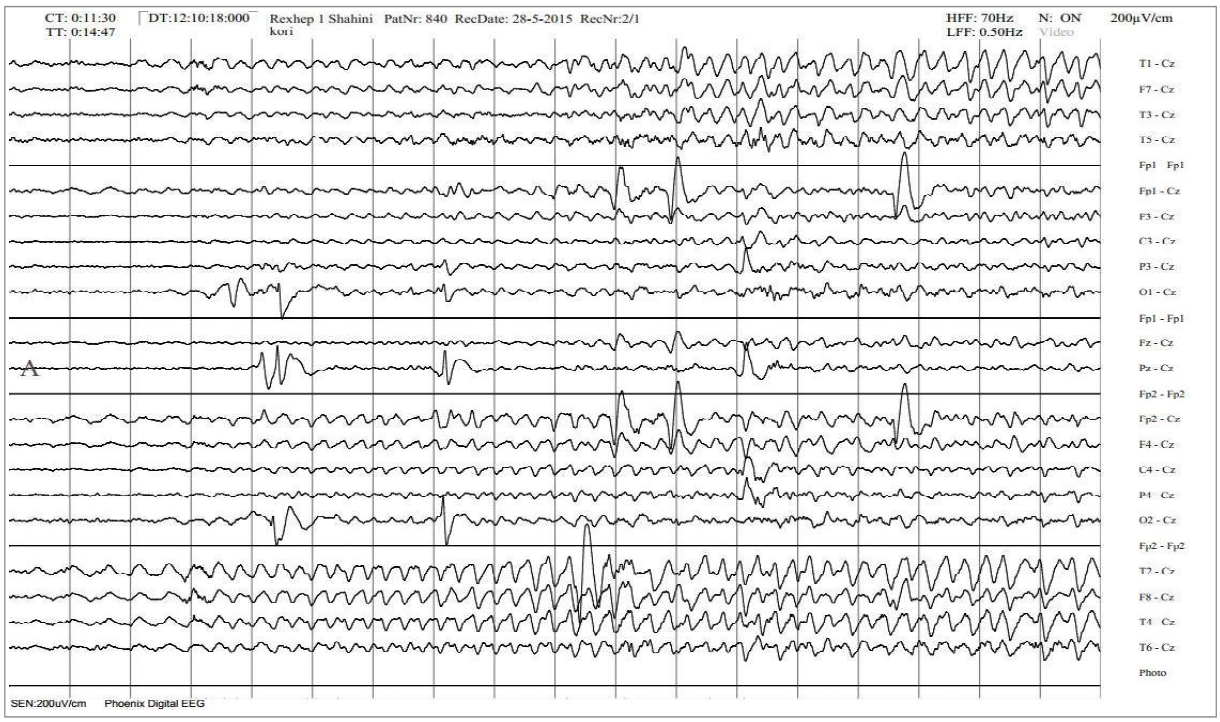
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Video
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T1 - Cz
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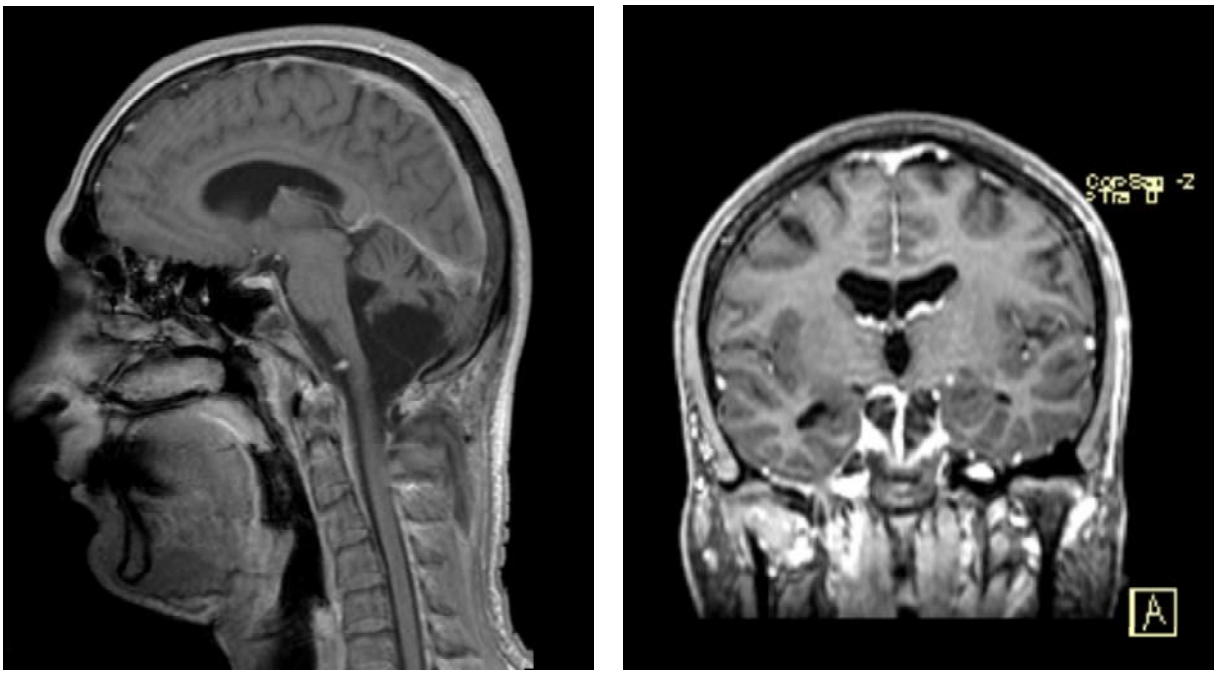
MRI imaging with epilepsy protocol revealed atrophy of the right hippocampal formation, fornix asymmetry, and neuropsychological evaluation suggested non-dominant temporal lobe (right) involvement. These data were congruent and epilepsy surgery multidisciplinary conference suggested anterior temporal lobectomy as the best option for seizure freedom for this patient.

Figure 3. a) Postoperative shrinkage of the ventricles, remodeling of the cerebellum and no signs of compression of the medulla oblongata. b) Atrophy of the right hippocampus with secondary dilation of the ventricular horn suggesting hippocampal sclerosis



Discussion

Our case is interesting because our patient suffering from chronic epilepsy, presented with signs and symptoms of progressive intracranial hypertension caused by an arachnoid cyst in the posterior fossa that was managed successfully with surgical removal of the cyst wall. The possible cause of the patient's epilepsy was only hypothesized clinically, but no apparent structural cause was possible in preoperative imaging studies. Hippocampal sclerosis was evident only after the shrinkage of the ventricles some time after surgery. Electroencephalographic data correlated the side of hippocampal atrophy with the epileptogenic region. We suggest that in posterior fossa arachnoid cysts or in posterior fossa cystic lesions causing hydrocephalus and associated with partial epilepsy, a supratentorial cause should always be sought before linking epilepsy with a possible, albeit rare cerebellar focus.

Conflicts of interest: None declared.

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