SVOA Neurology

ISSN: 2753-9180

ScienceVolks Open Access

Case Report

Video-EEG Findings in a Pediatric Patient with Moya-Moya Disease

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***Received:** March 14, 2022 **Published:** April 27, 2022*

Abstract

Objective: To assess the usefulness of the electroencephalogram (EEG) as a support for the diagnosis of Moya-Moya disease in a pediatric patient. The most common symptom in pediatric age is transient ischemic attack produced in hyperventilation (HV).

Method: Study of a patient admitted for video-EEG monitoring (VEEG) in the Neurophysiology Unit of the Virgen de las Nieves University Hospital in Granada, with a bibliographic review in PubMed. 8-year-old patient with symptoms of focal seizures without loss of consciousness, with the semiology of left hypotonia and headache caused after HV. It is evaluated for a suspected diagnosis of an idiopathic epileptic seizure.

Results: VEEG is performed, showing two critical episodes associated with post-HV with right hemispheric slowing, compatible with the "re-build up" phenomenon Through EEG analysis, Moya-Moya disease was suspected and later confirmed by angiography.

Conclusion: EEG analysis is an appropriate predictive method of diagnosis and follow-up for pediatric Moya-Moya disease.

Significance: An EEG is a non-invasive, simple test that can be easily performed in the pediatric population. There are characteristic findings in the EEG such as the phenomenon of "re-build up" that occurs after HV and they help to secure a diagnosis.

***Keywords:** Moya-Moya disease, "re-build up" phenomenon, electroencephalogram, hyperventilation, pediatric.*

Highlights

- There are clearly pathognomonic findings on the EEG with pediatric Moya-Moya disease.
- To evoke the characteristic "re-build up" phenomenon, it is necessary to perform optimal hyperventilation.
- EEG after hyperventilation is an important tool for both diagnosis and postoperative follow-ups.

Introduction

Moya-Moya disease is an idiopathic cerebrovascular condition produced by a progressive narrowing or occlusion of the terminal portions of the internal carotid artery (ICA) or proximal portion of the anterior cerebral (ACA) and/or middle cerebral (ACM) arteries. (Ann Cho et al, 2013)

The disease was first described in 1957 in Japan by Takeuchi and Shimizu. Its name comes from the Japanese word moyamoya: (もやもや), which refers to the "puff of smoke" appearance of the fine network of compensatory collateral vessels seen on angiography. (Gosalakkal, 2002)

Although its etiology is unknown, it is related to genetic alterations, found in genes such as RNF213, especially in the East Asian population. (Kim J, 2015) The incidence in Europe is approximately one-tenth of that observed in Japan. The association of this disease with some other primary entity makes it a syndrome (Dlamini et al., 2009), the causes of which include neurofibromatosis type 1 (NF-1), Down syndrome, Graves' disease, radiation therapy, and sickle cell anemia, among others. (Phi et al, 2015)

In epidemiological studies Moya-Moya disease affects a male-female ratio of 1: 1.8 or 1: 2.2, it occurs in two peaks, in childhood and between 35-50 years and approximately 10-15% of patients have family background. (Kim J, 2015)

The most common clinical symptoms in pediatric age are those associated with cerebral ischemia while in adults with cerebral hemorrhage (Gosalakkal J, 2002). Regarding the former, the most common is a transient ischemic attack (TIA), manifesting as motor alterations (70 to 80%) or as epileptic seizures (20-30%). (Kim et al., 2005) It is frequently associated with hyperventilation, with changes in temperature and situations such as crying or spicy food. (Ann Cho et al, 2013) Other symptoms such as headaches, aphasia, incoherent speech, hypotonia, etc. are also noticeably present. (Kacinski et al, 2007)

The results typical of pediatric patients with Moya-Moya in the EEG consist of slow waves with an increase in amplitude called posterior or centrotemporal slowing and the phenomenon of "re-build up", which consists of the asymmetric reappearance of slow waves (delta frequency) polymorphic with high amplitude (greater than 100 μV), 20-60 seconds after hyperventilation and with a duration of at least 30 seconds. For optimal stimulation, it is necessary to triple the expiratory volume at rest and last for more than 3 minutes. (Ann Cho et al, 2013)

Although there is no clear understanding of the "re-build up" phenomenon, it is believed to be produced by a decrease in cerebral perfusion causing regional cerebral hypoxia and a metabolic alteration of oxygen. It has been predominantly located deep in the cortical sulci. (Kacinski et al, 2007) This phenomenon disappears with age and it is rare to observe it in patients older than 13 years. (Ann Cho et al, 2013)

Ischemic damage may not progress or may progress insidiously, so surgical treatment can prevent brain damage and psychomotor impairment. (Kacinski et al, 2007)

Objective

To assess the usefulness of the EEG as a support for the diagnosis of Moya-Moya disease in a pediatric patient.

Method

The patient admitted to the Video-EEG Unit of the Virgen de las Nieves University Hospital in Granada (HUVN) on 12/3/2019. The VEEG is performed according to the International 10-20 system with a cap of electrodes and supernumerary sphenoidal and zygomatic electrodes, as well as an electrocardiogram, with an average and longitudinal bipolar assembly, standard impedances maintaining less than 5 Kilohms, low filter at 0.3 Hz, and high filter at 70 Hz, with sensitivity at 7 uV / mm and recording at 30 mm / second. These studies include stimulation tests through hyperventilation, which last 4 minutes and are performed at a rate of 30 breaths/minute.

The study lasts 10 hours. At the beginning of the registry, the patient was completely deprived of nocturnal sleep and without medication withdrawal.

The clinical semiology of the patient is analyzed together with the results of the VEEG tests and angiography. A bibliographic review was carried out in PubMed from 2002 to 2020.

Case

8-year-old female pediatric patient. She has a family history from her mother, siblings (17 and 15 years old), and several maternal relatives with migraines accompanied by paraesthesia and hemiparesis, triggered by exercise. She has a psychomotor development within normality and schooling without the need for support.

Patient with symptoms of focal seizures, without loss of consciousness, with the semiology of left hypotonia and headaches caused after hyperventilation. She had nonspecific headaches from the onset of the seizures. It is evaluated for a suspected diagnosis of an idiopathic epileptic seizure.

The crisis began when she was 5 years old with an abrupt start and end. The current frequency is less than 1 episode/ month with a duration 6-7 minutes. Episodes are always associated with hyperventilation during physical activity, intense and prolonged crying, or playing the flute, as well as changes in temperature.

The treatment is Oxcarbazepine: 450 - 0 - 450 mg (33 mg / kg)

She has the following complementary study:

- EEG (2017) - Performed in another Hospital Center: Hyperventilation causes generalized and symmetric slowing down of the tracing of physiological characteristics. During the procedure, the patient manifests dysesthesia and paresis in left arm, which fluctuates according to the indications of the explorer. During post hyperventilation, slowing is maintained in the hypervolted delta range with a clear predominance of the right hemisphere. In summary, EEG recording without intercritical activity and clinical crisis like the usual ones during hyperpnea, with sustained slowing during the post hyperventilation period.

Referred to the VEEG Neurophysiology Unit of the HUVN for study and diagnosis affiliation due to suspected focal seizures with motor and left sensory semiology.

Video EEG monitoring is performed on 12/3/2019 with a duration of 10 hours. In which waking periods, superficial (N1, N2) and deep (N3) NREM sleep phases are recorded. A basic activity is organized, symmetric, synchronous and reactive to stimuli. Background activity is not interrupted by the appearance of epileptiform activity.

Numerous stimulations are performed by hyperventilation with elevation of both arms. The hyperventilation performed induce a diffuse slowing of the tracing, predominantly in the left hemisphere. (Figure 1)

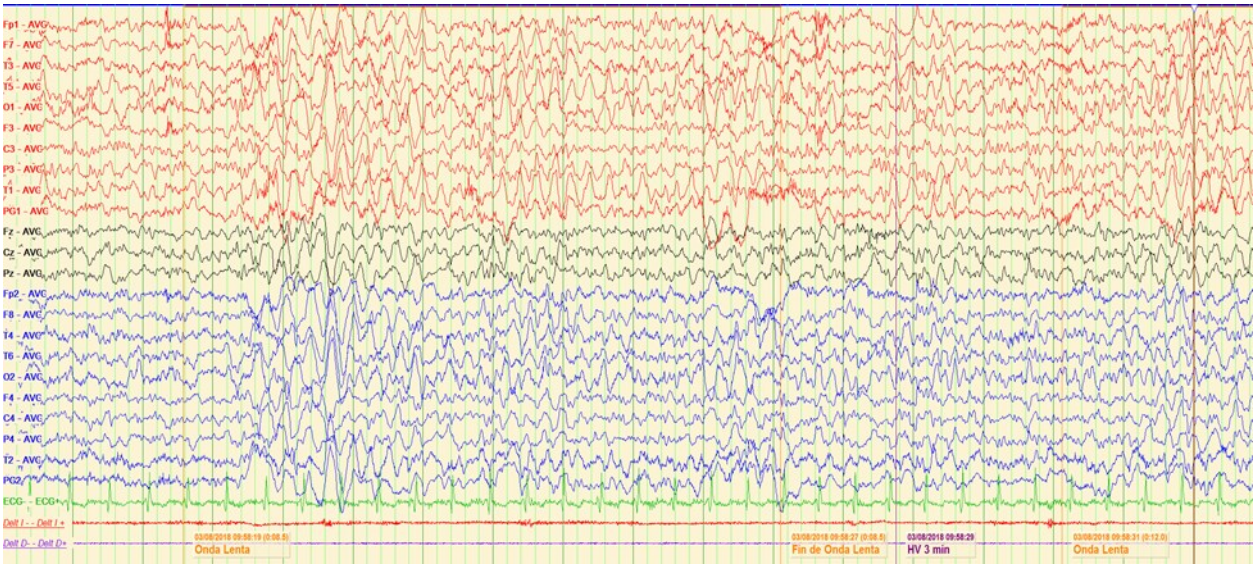

Figure 1: *Hyperventilation after 3 minutes.*

During the recording, 2 clinical episodes occurred in post hyperventilation without a decrease in the level of consciousness:

1ª Episode. Left arm paresis episode begins two minutes after the onset of focal slowing of the tracing.

EEG: Right hemispheric slowing in theta-delta range. 8.30 minutes duration that appears approximately 40 seconds after the hyperventilation. (Figure 2)

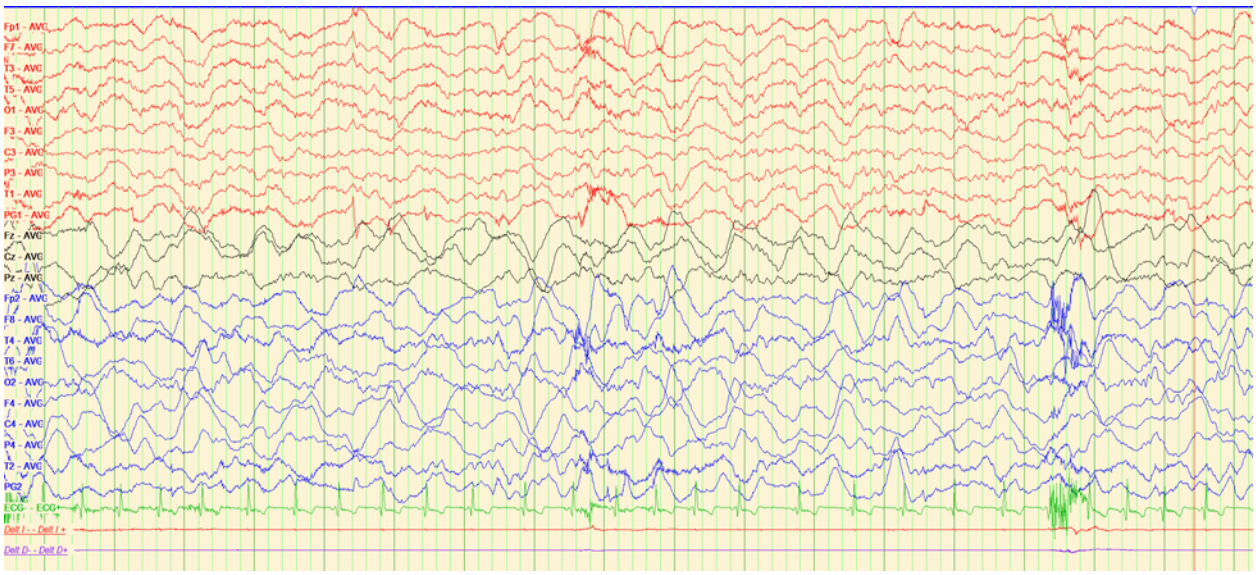

Figure 2: *Right hemispheric slowing in theta-delta range. 40 seconds after hyperventilation.*

2ª Episode. Left arm paresis and hypoesthesia followed by headache three minutes after the onset of the slowing of the tracing.

EEG: Irregular delta slowing of right frontal start, electrodes (Fp2, F8, PG2), which diffuses to the rest of the areas of the right hemisphere. 6:30 minutes duration. The slowing occurs approximately 1 minute after hyperventilation. (Figure 3)

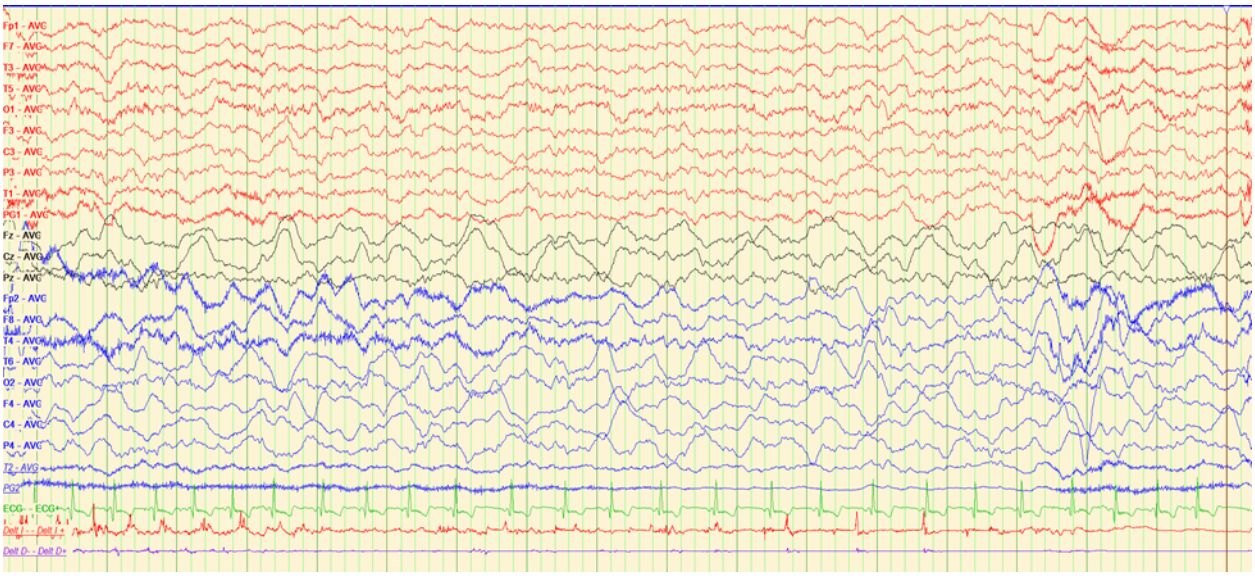

Figure 3: *Right hemispheric slowing in theta-delta range. 1 minute after hyperventilation.*

Recovery of left arm mobility and sensitivity is observed before the disappearance of the slowing of the traces.

These episodes are not electroencephalographically correlated with epileptiform activity. These crises are confirmed by a family member as like the usual ones.

In conclusion, Video-EEG study showing a diffuse slowing of the right hemisphere in post hyperventilation without appreciating intercritical or critical activity. During the recording, 2 episodes of paresis and paresthesia of the left arm occurred, both in post hyperventilation in relation to the right hemisphere slowing, compatible with the "re-build up" phenomenon.

Subsequently, on 8/13/2020, angiography was performed to confirm the suspected diagnosis:

• MRI-angiography of supra-aortic trunks and circle of Willis. Internal Carotids with progressive thinning of the supraclinoid segment on the right, complete disappearance of its middle and anterior branches and practically complete disappearance of the left anterior cerebral artery**.** (Figure 4)

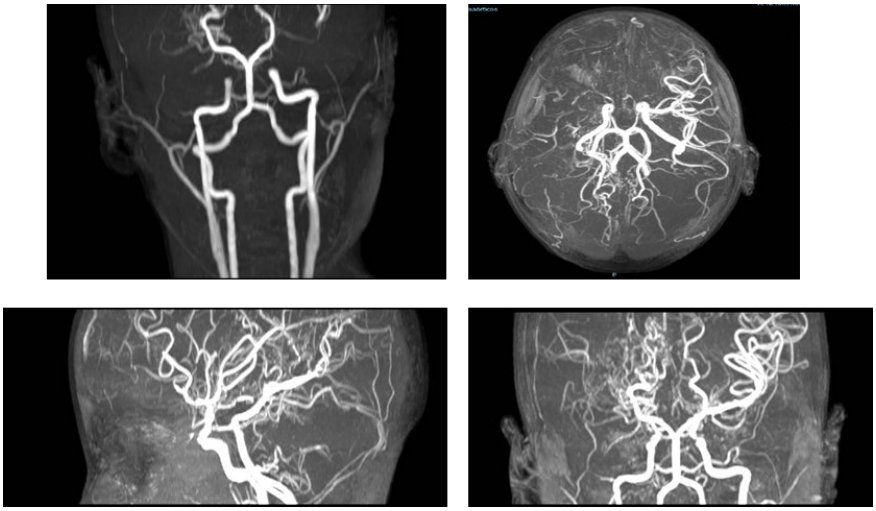

Figure 4: *MRI-angiography of supra-aortic trunks and circle of Willis.*

Discussion

Despite the indications of the family history and the symptoms of the seizures produced on all occasions after hyperventilation, the patient had a delay in the diagnosis. A possible migraine origin of the episodes was considered due to its association with some of them, since Moya-Moya disease, which is rare in the European population, was not considered at first (Yáñez et al., 2008). She started the clinic at 5 years of age, which had little interference with her daily life, and was admitted to the VEEG monitoring unit until she was 8 years old. Through the VEEG analysis, Moya-Moya disease was suspected and later confirmed with an angiography.

The diagnostic criteria establish that, in the case of unilateral involvement, catheter angiography is necessary, while in bilateral cases magnetic resonance imaging/angiography can be used. (Kim J, 2015) An EEG is a non-invasive, simple test that can be easily performed unlike an angiography in the pediatric population.

There are clearly pathognomonic findings on the EEG with pediatric Moya-Moya disease. (Qiao et al., 2003) To evoke the characteristic “re-build up” phenomenon, it is necessary to perform optimal hyperventilation. Evaluation of the EEG after hyperventilation is an important tool for both diagnosis and postoperative follow-up. (Kacinski et al, 2007)

The phenomenon of “re-build up” disappears after an effective bypass intervention (Qiao et al., 2003) Therefore, the EEG, being a simple specific indicator of the alteration of cerebral hemodynamics, allows monitoring patients after the intervention.

However, there are reviews that advise against performing such stimulation despite the fact that others emphasize the need to perform it for diagnosis and follow-up. (Dlamini et al., 2009) This discrepancy could be the cause of the few studies that address the clinical utility between EEG and Moya-Moya disease due to lack of consideration.

In reference to this, Ann Cho et al, establish in their study carried out in 2013 that hyperventilation in the EEG can be used as a relatively safe and evaluation tool of the cerebral vascular reserve without causing any significant neurological deficit, always being carried out under the supervision of qualified personnel and asking the patient to stop when they notice symptoms. Before admission, the patient had two EEGs performed where a sufficiently optimal hyperventilation was not performed as the one carried out in the VEEG that produced the “re-build up” phenomenon.

In pediatric patients, a predominant perfusion deficit has been observed after hyperventilation in the frontal lobe. Coinciding with this, the “re-build up” phenomenon is mainly located deep in the grooves of the cortex with less perfusion. (Kacinski et al., 2009). The case-patient, like most of the patients reviewed in the literature, presents the phenomenon of “re-build up” in the frontal area.

The risk of suffering from it in family members is 30-40 times higher than in the general population (Kim J, 2015); Given their hereditary nature and the symptoms that the mother and the patient's siblings present, they undergo MRI angiography to screen for Moya-Moya disease. Although conventional angiography is the gold standard in diagnosis, it has its relative limitations for follow-up, due to its invasive nature and possible complications. (Ann Cho et al, 2013) It would be advisable after the patient's intervention, an EEG will be performed with optimal hyperventilation, to objectify the disappearance of the “re-build up” phenomenon that allows providing data to assess the evolution in a simple way.

Conclusion

Analysis through the EEG is an appropriate predictive method of diagnosis and follow-up for pediatric Moya-Moya disease.

Conflict of Interest

None of the authors have potential conflicts of interest to be disclosed.

Authors contribution

Galdón Castillo conceived of the presented idea. Ortega León acquired the data. Ruiz Serrano L and Ruiz Navarrete review literature and discussed the results. All authors contributed to the final manuscript.

Acknowledgement

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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Citation: Ruiz Navarrete PA, Ruiz Serrano L, León Ortega MT, Galdón Castillo A. "Video-EEG Findings in a Pediatric Patient with Moya-Moya Disease". SVOA Neurology 3:3 (2022) Pages 85-90.