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CASE REPORT

A rare concurrence: gelastic seizures in a patient with right temporal nongalenic pial arteriovenous fistula

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Abstract

Introduction Gelastic seizures are the type of seizures that are most commonly seen in childhood and should be excluded definitely in the differential diagnosis of hypothalamic hamartomas. This seizure type may be accompanied by refractory seizures, cognitive decline, and early puberty. However, etiology may also include other causes different than hypothalamic hamartomas. The seizure may also arise from temporal and frontal region, in addition to hypothalamus. Different clinical findings may be observed based on origin and areas of spread.

Conclusions In this article, we report a case of gelastic seizure that has been observed by a different cause other than hypothalamic hamartoma which was reported for the first time in the literature.

Keywords Arteriovenous fistula · Gelastic seizure · Hypothalamic hamartomas · Temporal region

Introduction

Gelastic seizures (GSs) are the type of seizures that are quite different from other types of seizures that usually arise from hypothalamic hamartomas (HH). It was first described in 1868 by Trousseau in a boy with meaningless laughter attacks but unconscious of it as well as no remembering at all, and it was stated that this might be a seizure [1]. In 1957, Daly and Mulder used the term "gelastic seizure" for the first time [2]. In 1971, Gascon and Lombroso defined the characteristics of GSs as: "stereotypic, repetitive, out of social context laughter attacks, which may be related to ictal or interictal discharges on the electroencephelogram (EEG), or may be associated with other types of seizures" [3].

GSs are rare type of seizures arising from hypothalamus, most commonly seen in children and most commonly related to HHs. These seizures may be accompanied by pubertal disorders, cognitive decline, and refractory seizures. Even though, HHs should be excluded in such seizures. GSs can occur rarely due to different causes seen more frequently in adults [4–6]. We report a case of pediatric GS due to a different cause other than HH. To the best of our knowledge, this case is the first pediatric one reported in the literature.

Case

A healthy 6-year-old boy presented to our clinic with complaints of sudden eye deviation lasting 2–3 s after laughing, followed by left hand and foot weakness over a month. It was stated that laughter was out of social context, sudden, full of joy, accompanied by loss of consciousness, and then weakness developed. Initially, this has happened once a day, but has recently started to be 20–30 times a day. There was no important feature in his history. His family history was noncontributory. The patient's school success was good and his neurological examination was normal. There was no abnormality in routine laboratory examinations of the patient. On the EEG, there were spike and wave discharges at right centrotemporal region (Fig. 1). Cranial magnetic resonance imaging (MRI) showed arteriovenous fistula in the right temporal lobe (Fig. 2), then it was detected that the pathogenic variant at the magnetic resonance angiography was nongalenic pial

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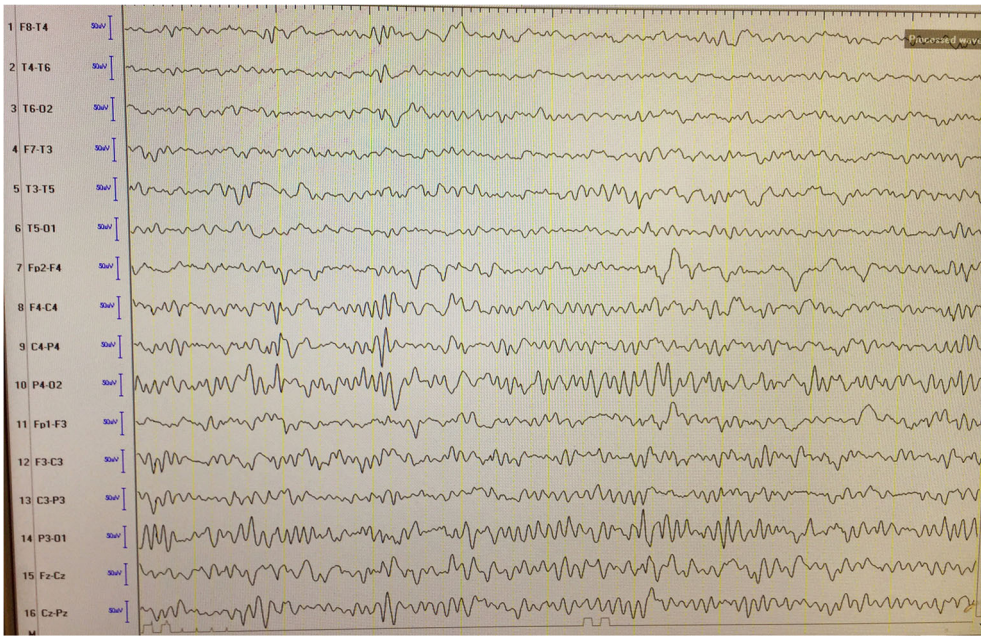

Fig. 1 Spike and wave discharges at the right centrotemporal region

arteriovenous fistula (NGPAVF) (Fig. 3). The NGPAVF was occluded with endovascular coil embolization by digital subtraction angiography (DSA) (Fig. 4). The seizures were controlled by levetiracetam and the patient has been seizure free for 2 years.

Discussion

GSs are very rare type of seizures. It is an epileptic syndrome which is usually seen in children, in which HHs are involved in etiology and which can be accompanied by puberteprecox, refractory seizures, and cognitive decline. Tumors, tuberous sclerosis, cortical malformations, and post infectious focus are present in the etiology especially in adulthood except HH [7]. To the best of our knowledge, the NGPAVF is the first case related to GS reported in the literature.

GSs can be raised not only from the hypothalamus but also from the temporal and frontal lobes. Connections are provided with hypothalamus via networks formed in these regions. Findings vary in relation to which part of the brain the seizure originates [8] such as laughing with joy, loss of consciousness, aura, accompanied by other types of seizures (for example, atypical absences, atonic seizures, automotor seizures, hypomotor seizures, generalized tonic clonic seizures). Dericioglu et al. [8] reported that the seizures especially raised by the temporal lobes were spread to the basal temporal region and joyful laughter occurred as a result. Coria et al. [9] presented a GS case with joyful laughter, originating from the right temporal region. Kovac et al. [7] reported that aura, loss

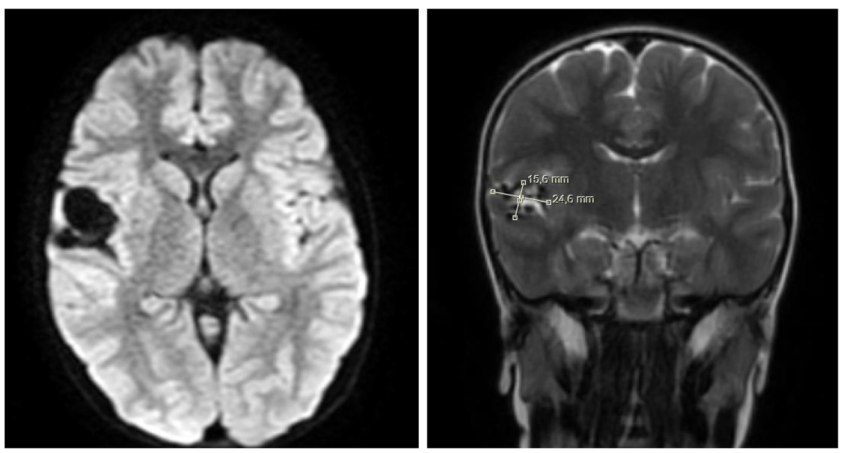

Fig. 2 T2-weighted sequence. Axial and coronal view showing nongalenic pial arteriovenous fistula at right temporal region

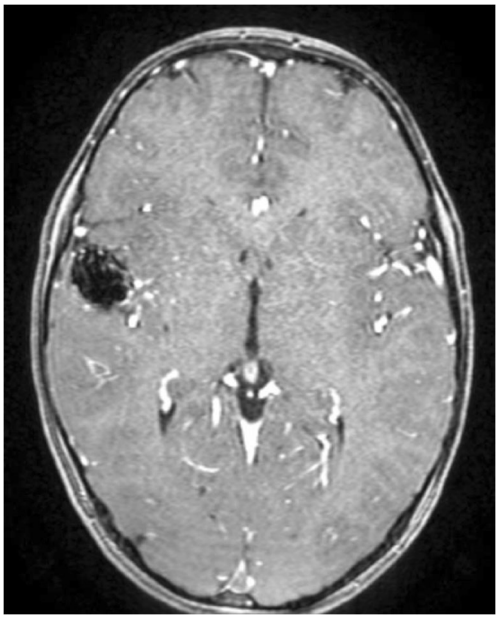

Fig. 3 MR angiography view showing nongalenic pial arteriovenous fistula at right temporal region

of consciousness, and automotor seizure different than laughing joyfully may be accompanied by GS raised from the temporal region. Even though, it is stated that GSs involved from frontal region are more emotion free and contain much more motor component.

In our case, the NGPAVF is present in the right temporal lobe and it is thought that the seizure is caused by this region. The NGPAVF is a direct connection between intracranial artery and vein without nidus. Clinical manifestations of the NGPAVF are based on the steal phenomenon due to high-velocity arteriovenous shunt and its mass effect [10]. The clinical findings can be explained to the cerebral ischemia due to the steal blood. In our case, the NGPAVF was located at right temporal region and it stole blood from right MCA, M3 segment. As known, the MCA, M3 segment feeds to insular, frontoparietal operculum and sylvian fissure regions. We consider that all of the clinical manifestations about patient occur due to reduced blood flow from these regions.

Also, it has been shown that GS arise from the anterior cingulate cortex, premotor frontal cortex, and operculoinsular areas through projections extending to the motor cortex and ventral brainstem [11, 12]. In our case, there was GS with joyful laughter, loss of consciousness, and after Todd's paralysis clinic. After the arteriovenous fistula was occluded by radiosurgery, the seizures were controlled by levetiracetam and the patient is now seizure free for 2 years. In addition, since the patient's seizures were stopped after endovascular coil embolization, so it was thought that the etiology of this seizure was effective on the steal of blood instead of mass effect. Currently, the treatment of the NGPAVF is surgical or endovascular embolization [10]. Embolization was preferred because of complications related to surgery and postoperative neurologic complications, the risk of hemorrhage.

In conclusion, it is known that GS develops due to HH in childhood and it is associated with puberty precox, cognitive decline, and refractory seizures. However, our case is different than the literature from the following points: the GS occurred with the lack of HH, lack of cognitive decline, and the control of seizures with a single antiepileptic drug. To the best of our knowledge, the NGPAVF is the first case reported in the literature in GS etiology.

Compliance with ethical standards

Conflict of interest The authors declare that there is no conflict of interest.

Ethics approval and consent to participate The presentation of the case was approved by the Clinical Research Ethics Committee of the Mersin University. Informed consent was obtained.

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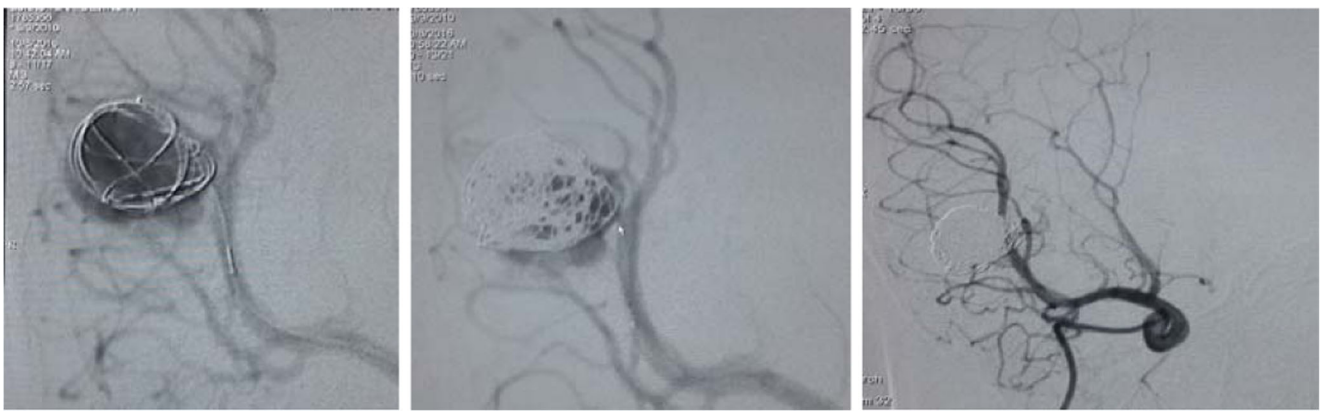

Fig. 4 Digital subtraction angiography (DSA) view showing nongalenic pial arteriovenous fistula at right temporal region

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