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Hot Water Epilepsy: Presentation of Three Cases

Sıcak Su Epilepsisi: Üç Olgunun Sunumu

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Summary

Hot water epilepsy (HWE) is a reflex epilepsy that develops after pouring hot water on the head; seizures are induced through tactile and temperature-related stimuli. The number of cases reported worldwide is low, with most cases in Turkey and India. The exact pathophysiology of HWE is unknown but patients are thought to have abnormal thermoregulation systems with seizures that emerge due to the stimulation of a particular region in the brain cortex via contact of hot water on the skin of the head. We investigated the pathogenesis of this disorder through a literature review and by presenting the clinical and laboratory findings of three patients with HWE. Fortunately, HWE can largely be prevented; however, if non-reflexive seizures co-occur with HWE, proper medical treatment can be added to ensure seizure-free follow-up.

Keywords: Hot water, epilepsy, reflex epilepsy

Öz

Sıcak suyun baştan aşağı dökülmesi ile oluşan, taktil ve sıcaklığa bağlı stimuluslarla uyarılan nöbetlerin ortaya çıktığı refleks epilepsi tipi sıcak su epilepsisi (SSE) olarak tanımlanmıştır. Literatürde en çok Türkiye ve Hindistan'dan olgu bildirilmesine rağmen tüm dünyada bildirilmiş olgu sayısı azdır. SSE'nin patofizyolojisi tam olarak bilinmemektedir, ancak hastaların anormal bir termoregülasyon sistemlerinin olduğu, kafa derisinin sıcak suyla teması ile beyin korteksindeki belirli bir bölgenin uyarılması nedeniyle nöbetlerin ortaya çıktığı düşünülmektedir. Çalışmamızda, SSE olan üç hastanın klinik ve laboratuvar bulguları sunularak hastalığın patogenezi literatür eşliğinde incelenmiştir. SSE, gerekli önlemler alındığında çoğunlukla önlenebilen bir refleks epilepsi tipidir. Ancak hastalarda SSE ile birlikte non-refleksif nöbetler de eşlik ediyorsa, uygun medikal tedavi alınan önlemlere eklenerek nöbetsiz başarılı takip sürelerine ulaşılabilir.

Anahtar Kelimeler: Sıcak su, epilepsisi, refleks epilepsi

Introduction

Epilepsy that emerges as a response to specific stimuli such as flashing lights, visual stimuli, eating, and listening to music is called "reflex epilepsy". Reflex epilepsy is rare and constitutes about 6% of all epilepsy cases (1). The type of reflex epilepsy that develops after pouring hot water on the head and seizures are induced by tactile and temperature-related stimuli of water are defined as hot water epilepsy (HWE) (2). HWE constitutes 3.6-3.9% of all epilepsy cases in India, and most reported cases are from India (3). HWE is most commonly seen during childhood and in males (4). The seizure type can be generalized as tonic clonic as well as complex partial (5). Here, we investigate the pathogenesis of the disorder through a literature review and by

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presenting the clinical and laboratory findings of three patients with HWE.

Case Reports

Case 1

A boy aged 3 years described contractions with resulting bruising over the whole body when water was poured on his head for the last two months. The prenatal, natal, and postnatal history revealed nothing significant. There was no febrile convulsion and no family history of epilepsy. The parents were not consanguineous. The contractions emerged within one minute after hot water was poured on the head during bathing and continued for about 2-3 minutes. Motor and mental development and other system findings were normal. The hemogram, biochemistry results, urinalysis, and electrocardiogram (ECG) were normal. Cerebral magnetic resonance imaging (MRI) results were also normal. The routine awake and sleep electroencephalograms (EEG) obtained in the interictal period were also normal. The patient was diagnosed as having HWE. Decreasing the bath temperature was recommended for treatment, but the seizures emerged even with low bath temperature, warm water or little amount of water. The patient was therefore prescribed 0.01 mg/kg clonazepam to be taken before bathing. The patient had no seizures for the next six months of follow-up once treatment was started.

Case 2

A boy aged 6 years presented with symptoms of bruising, deviation of the eyes, and full-body contractions after contact with hot both water, which he had had since the age of 3 years. The prenatal, natal, and postnatal history revealed nothing of significance. There was no febrile convulsion and no family history of epilepsy. The parents were not consanguineous. The symptoms were found to last for a couple of minutes, and he completely regained consciousness afterwards. The symptoms had been observed when pouring hot water, especially to the head, during almost every bath. The family stated that he did not experience seizures when bathing with warm water. Motor and mental development were normal. The hemogram, biochemistry results, urinalysis, and ECG were normal. Cranial MRI and interictal EEG were normal. The patient was followed-up after being diagnosed as having HWE. We recommended lowering the bath temperature and to wash the head region with less water at low temperature. The patient had no further seizures in the 2 years of follow-up.

Case 3

A girl aged 7 years presented with a history of generalized tonic clonic seizures that had started 2 years ago and had emerged during every bath in the last year. The prenatal, natal, and postnatal history revealed nothing of significance. There was no family history of epilepsy. The parents were not consanguineous. The seizures were found to last for 1-2 minutes. The seizure frequency and duration had recently increased. Psychomotor development, as well as the neuromuscular system and systemic exam were normal. The hemogram, biochemistry results, urinalysis, and ECG were normal. Cerebral MRI and interictal EEG were normal. The patient had not been treated before and was recommended to bathe in tepid water and to wash the head region in particular with lukewarm water only. The seizures continued despite the measures taken so she was put on valproate. The patient did not experience seizures in the 6 months after valproate treatment was started.

Discussion

HWE was first identified in 1945 in New Zealand by Allen (1). The largest case series in the literature was reported from Turkey and India, but the number of reported cases is still low (2). The definite pathophysiology of HWE is not known but these patients are thought to have an abnormal thermoregulation system with the seizures emerging due to the stimulation of a particular region in the brain cortex through contact of head skin with hot water (6). The cause of this abnormal thermoregulation system is thought to be an autosomal recessive hereditary predisposition; this hypothesis has been supported by autopsy studies (7,8). Although consanguineous marriages are common in Turkey and especially in the Kars region, there was no kinship between the parents in these cases. The water temperature that causes seizures ranges between 40-50 ºC (7). However, water temperature is not the only factor that causes seizures; there are many differences between HWE patients. Factors such as the body area where the water is applied, bathing duration, and bathing style (showering or pouring water on the head) are important factors in the emergence of HWE symptoms. Water at 50-55 ºC has been found to cause epileptic seizures within 3.5 to 10 minutes (9).

MRI and EEG are usually normal in patients with HWE. Hyperperfusion areas have been reported via single photon-emission computed tomography in certain cases of HWE (10). Patients with HWE who have focal epileptic activity originating from the temporal or parietal lobes on EEG, temporal hippocampal sclerosis, dysplasia, cystic changes, and other abnormal findings such as cavum septopellucidum and pachygyria on MRI have also been reported (4,11,12,13). The MRI and EEG results in our patients were normal. This is consistent with the literature.

The types of seizure that occur in patients with HWE are complex partial (67%) and generalized tonic-clonic (33%) (14). All of our patients experienced generalized tonic clonic seizures. HWE is usually seen in children and various studies have evaluated the seizure onset age. The mean seizure onset age was 13.4 years in the largest series in the literature, but it can be as low as 4.7 years (14,15). We found that the first seizures developed at a mean age of 3.6 years in our patients. We believe that the mean age of the first HWE seizure occurrence seen here might be lower than that reported in the literature due to our low number of cases. High water temperatures are preferred while bathing children due to the cold weather in the Kars province located in the Eastern Anatolia region of Turkey, which has a continental climate. Thus, we recommended using lower water temperatures and a decreased volume of water. No new seizures were seen during approximately 2 years of follow-up with this method. However, about one third of patients with HWE continue to have seizures even with lukewarm baths. These patients develop spontaneous seizures during follow-up (3,7,16). It was reported that spontaneous seizures can occur at rates between 16-62% (5,7). Conventional epileptic drugs are often used as medical treatment similar to our protocol. Prospective studies have also recommended intermittent oral clobazam as a prophylactic treatment before bathing (17). We

also facilitated seizure-free periods with clonazepam in one patient and valproate in another.

In conclusion, HWE is a type of reflex epilepsy that can generally be prevented when the necessary measures are taken. However, proper medical treatment can be added to these measures to ensure seizure-free follow-up if non-reflexive seizures are also present.

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: Yusuf Ehi, Sefer Üstebay, Döndü Ülker Üstebay, İnan Gezgin, Şeyho Cem Yücetaş, Design: Yusuf Ehi, Sefer Üstebay, Döndü Ülker Üstebay, Data Collection or Processing: Yusuf Ehi, İnan Gezgin, Şeyho Cem Yücetaş, Analysis or Interpretation: Yusuf Ehi, Literature Search: Yusuf Ehi, Writing: Yusuf Ehi.

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