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Non-epileptic attack disorder: the importance of diagnosis and treatment

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TITLE OF CASE

Non-epileptic attack disorder: the importance of diagnosis and treatment

SUMMARY

A fifty-year old woman was taken to hospital by emergency ambulance during her first seizure. She was admitted to hospital, treated with IV diazepam, diagnosed with epilepsy and started on anti-epileptic drug (AED) therapy. This was ineffective so she was referred to a tertiary centre where she underwent video EEG and was diagnosed with non-epileptic attack disorder. Her experience of the diagnosis was positive, it allowed her to understand what was happening to her and to understand the link between her seizures, adverse childhood experiences and the death of her mother. She stopped taking AEDs and she was referred to a psychologist which led to a significant improvement in her functioning and quality of life. We present this case as a good example of the benefits of accurate diagnosis, clear explanation and access to specialist care.

BACKGROUND

Non-epileptic attack disorder (NEAD) is characterised by episodic disturbances of normal function and control that superficially resemble epileptic attacks but are not caused by epileptic activity in the brain and are thought to have a psychological basis [1] [2]. Diagnosing NEAD can be difficult even for specialists but it can reliably be distinguished from epilepsy using video-EEG which shows normal electrical activity in the brain during attacks. Most patients with NEAD currently receive an initial diagnosis of epilepsy and there is often a delay of several years between the initial seizure manifestations and eventual diagnosis of NEAD [3]. Misdiagnosis leads to emotional distress, confusion, inappropriate use of anti-epileptic medication, repeated attendance at emergency departments (EDs) and prevents access to psychological treatments [4]. Prolonged seizures are often misdiagnosed in EDs as status epilepticus leading to inappropriate treatment with intravenous benzodiazepines, anaesthesia and admission to ITU, putting patients at risk of the adverse consequences of these treatments that include death. Patients often experience negative attitudes from health care professionals, many of whom misunderstand NEAD, and believe that patients are consciously faking epileptic seizures during their attacks.

CASE PRESENTATION

Medical History

A fifty-year old woman started having almost daily seizures in October 2011. During her first seizure she was taken to hospital by emergency ambulance, treated with IV diazepam and admitted to hospital. She remained an inpatient on a medical ward for ten weeks because she continued to have regular seizures despite AED therapy. The seizures were stereotyped and lasted up to one hour, they were preceded by a metallic taste, and involved generalised limb jerking, frothing from the mouth, incontinence and tongue biting (see Video for a typical seizure). Her medical history included fibromyalgia, migraine, chronic daily headache, depression and anxiety (mild), osteoporosis, tonsillectomy, appendicectomy, laminectomy, hysterectomy, cholecystectomy and head injury (she was assaulted in 1993). After discharge from hospital in December, she continued to have seizures every few weeks despite treatment with sodium valproate, carbamazepine, gabapentin and topiramate (topiramate was prescribed mainly for treatment of migraines) so she was referred to a tertiary epilepsy centre for further assessment.

Social and Family History

She was the youngest and only girl in a family of six children. Her father sexually abused her from the age of five with the abuse continuing into her adult life until her father's death.

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She was the only sibling that was sexually abused. However, one of her nieces was also abused by her father, and there was a strong suspicion that he had abused a second niece, and a suspicion of abuse of others outside the immediate family. Some of the abusive incidents took place in public places and in front of others. She had understood her father's life to have been very hard, with early life rejection and abuse including broken limbs. Her father was verbally abusive to her mother and physically violent to her brothers, all of whom had turbulent relationships with their father as adults. She had a close relationship with her mother who died in September 2010. She suffered bereavements in 2012 when her nephew died quite suddenly of a brain tumour at the age of 35, and in 2014 when her brother (the father of her nephew) was killed in a road traffic accident whilst abroad. She was very concerned about the care that her mother received during her final illness and very upset to be informed of her death over the telephone. This led to a long and stressful dispute with the hospital that eventually acknowledged significant failings. She has no children and is happily married to a very loving and supportive husband.

INVESTIGATIONS

MRI showed deep white matter high signal intensities on the T2-weighted flair images consistent with moderate vascular disease and an area of cortical gliosis in the right frontal lobe. The MRI scan was otherwise normal. She had continuous 48-hour video telemetry on 03/10/12 during which she had a typical attack. It began with a metallic taste, she lowered herself to the floor and after 15 seconds generalised asynchronous shaking of her limbs and trunk began. The seizure lasted approximately 6-7 minutes. She was unresponsive throughout and intermittently responsive afterwards. She slept for a short period afterwards but made a quick recovery. Her ictal and inter-ictal EEGs throughout the telemetry showed no evidence of epileptic activity.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of suspected seizures is long but over 90% of self-limiting episodes of unprovoked transient loss of consciousness (TLOC) are caused by epileptic seizures, vasovagal syncope and NEAD [2]. Cardiovascular conditions which cause TLOC are often associated with brief myoclonic jerks that can be mistaken for epileptic seizures. Vasovagal syncope is the most common cardiovascular cause of TLOC but other potentially serious conditions such as cardiac dysrhythmias can cause TLOC.

OUTCOME AND FOLLOW-UP

In October 2012, after review of the telemetry reports, she was diagnosed with NEAD (by MR) and her AEDs were stopped. She was provided with information about NEAD, with sources of explanation and support (including www.nonepilepticattacks.info) and follow-up was arranged. The delivery of a diagnosis which clarified that the condition was "real" and where the seizures were understood to be "her mind shutting down" in response to experiences of trauma, stress and bereavement, was very helpful to her. She described the communication of the diagnosis as a turning point in her life which enabled her to make sense of and better manage her seizures. It is the loss of her mother and subsequent dispute with the hospital over her care that our patient and her husband believe were the most important aetiological factors in her seizures. After the diagnosis, she was less distressed when seizures occurred and she felt able to re-establish her previous activities sooner. After her first appointment she continued to be followed-up in clinic. Overall her seizure frequency and severity improved after the diagnosis and in the subsequent years. In 2014 her brother was killed and her father-in-law died which triggered an increase in frequency of her seizures. But by October 2015 daytime seizures had largely disappeared (2-3 per month of short duration) leaving her with mainly nocturnal seizures.

Ensuring that she received the most appropriate emergency care was difficult for the first

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few years of her illness when she was having frequent and often prolonged seizures many of which were complicated by facial injuries especially abrasions and lacerations which required cleaning, stitching and dressing. It took time for her husband to feel confident with managing short simple seizures without calling an emergency ambulance and then for him to develop confidence with longer seizures. Communicating the diagnosis during a seizure to paramedics and emergency doctors who had no prior personal knowledge of the case was also difficult especially in terms of avoiding inappropriate use of benzodiazepines. Within a few years of diagnosis an effective emergency care plan had been developed in collaboration with her neurologist and the A&E consultants in the local hospital which her husband was able share with paramedics and doctors when required. As a result of the improvement in her seizures she has not been taken to A&E or sustained any injuries since August 2015.

She was referred to a clinical psychologist with expertise in NEAD and she has been under her care on-and-off since 2013. There were long waits between referrals and appointments because a suitable service was not funded locally and requests for special funding were necessary. She declined psychological treatment for NEAD and because she had already undertaken extensive psychotherapy between 2007-2010 for her headaches and fibromyalgia where the links between her physical symptoms and childhood sexual abuse (CSA) were explored. During that time, she had been a member of a support group for victims of abuse and she didn't feel further focused psychological treatment would be helpful. Nevertheless, the psychologist was able to offer explanation about the diagnosis, support and counselling which the patient found very valuable.

DISCUSSION

There is increasing understanding of NEAD but much remains unknown. As yet there is no consensus on terminology, with non-epileptic attack disorder, psychogenic non-epileptic seizures (PNES), functional seizures, dissociative seizures all in current usage. The term pseudo-seizures is still sometimes used but is now regarded as out-dated and pejorative [1]. The lack of a simple label and a mechanistic account of the pathogenesis of the disorder as well as the stigma associated with seizures and mental health disorders make the explanation of the diagnosis problematic. Some patients are very resistant to the diagnosis and the idea that "it is all in my mind" can cause much upset and confusion for patients and doctors. The method of delivering the diagnosis of NEAD is important, some authors suggest that if done well it can result in reduction in seizure frequency, even cessation of seizures, and reduction in healthcare utilisation (especially the use of emergency services) [4] but negative experiences of diagnosis may worsen a patient's prognosis. Several protocols have been developed for delivering the diagnosis [5] [6] [7] but more research is required to establish their efficacy [8].

A common misunderstanding is that non-epileptic attacks are consciously staged or faked, that non-epileptic attacks are voluntary and under conscious control, and that they can be initiated or terminated at will. These misunderstandings are commonly held by healthcare professionals and are likely to give rise to overtly, or poorly concealed, hostile communication and behaviour, and low quality care [9]. The aetiology of NEAD is uncertain. Patients have high levels of somatisation, dissociation and general psychopathology and it seems that patients with NEAD have a tendency to manifest psychosocial distress as somatic symptoms [10]. Dissociation is negatively correlated with quality of life [11] and there is often a mismatch between subjective reports of anxiety and physical arousal [1]. Better understanding, better communication, good quality information provision [12] and education are essential to improve care for patients with NEAD.

The accurate and timely diagnosis of NEAD is a crucial step in providing the best quality care but the diagnosis is difficult to make so the seizures are often compounded by frustration and confusion on the part of the doctor and the patient [13]. The mean delay to diagnosis is seven years. Patients are often treated by non-specialists who lack detailed knowledge of NEAD and epilepsy. There are no useful investigations in the inter-ictal phase, the diagnosis

largely relies on the semiology and history but these are imperfect discriminators. Without an accurate diagnosis, or at least serious consideration of NEAD as part of the differential diagnosis, patients are put at risk of serious iatrogenic harm from emergency treatment especially benzodiazepine treatment, anaesthesia, ITU admission and their complications. In the longer term they may be unnecessarily exposed to the side-effects of AEDs [14] while access to psychological treatments which have been shown to be effective may not be considered [4]. The diagnosis of NEAD is typically made by specialised epilepsy services but the geographical coverage of these is variable with no access in some areas. Specialised psychotherapy has been shown to be effective, but these services are very limited (more generic CBT approaches to treatment may help).

Factors which have been shown to predispose to, precipitate and perpetuate NEAD are traumatic life events especially abuse (both sexual and non-sexual), bereavement and family illness. There can be long latencies between life-events and the onset of seizures, multiple events can interact and some life events seem to predispose to rather than cause non-epileptic attacks [15]. When making detailed enquiries about social history and life events from the patient and significant others, doctors may be concerned about causing distress or offense, they may be worried that it could give rise to complaints or even legal action and they may be concerned about the amount of time required [13] but the social history, and identification of risk factors for NEAD, are very important parts of the assessment and they should be actively, sensitively and respectfully explored by clinicians.

There is limited evidence on prognosis [16] [17] in NEAD. Most patients improve in the short/medium term but initial improvement does not rule out eventual relapse and a significant proportion have long-term intractable seizures. Although, the available data shows that long-term outcomes vary, some patients make a full recovery and the possibility of a good recovery should be made clear to patients at diagnosis and when discussing psychological treatment.

LEARNING POINTS/TAKE HOME MESSAGES

Accurate diagnosis and careful explanation of NEAD is important [18] [19]. It enables the patient to understand what is happening to them, avoids iatrogenic harm from inappropriate treatment and allows access to psychological therapies that can improve wellbeing.

NEAD should be considered in the differential diagnosis of seizure disorders at all stages in the diagnostic pathway including the first presentation.

The social history is an essential part of the assessment of patients with suspected seizures and it is appropriate that it should be actively, sensitively and respectfully explored by clinicians involved in their assessment.

Psychological therapy can improve the outcome of patients with NEAD and all patients should be offered access to this treatment.

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FIGURE/VIDEO CAPTIONS

Video of the patient experiencing a non-epileptic attack. The video was taken at home by the patient's husband.

PATIENT'S PERSPECTIVE

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The first draft of this case report was written by the authors. JMD and MP met the patient and her husband who gave consent for the report to be written and submitted and provided them with copies of the draft and invited them to give comments and to write their perspective in their own words. They approved the final manuscript and video, gave written consent to its publication and provided the following comments.

Patients' Perspective. Reference has been made to sexual abuse sustained throughout childhood and beyond. I feel that this was dealt with through a prolonged course of CBT. However later traumatic events perhaps pushed me over the top triggering NEAD. I am sure that the appalling treatment of my mother during her last 5 months, all spent in hospital, affected me greatly. The ensuing battle lasting 5 years with the hospital's complaints system only made that worse.

Husbands' perspective. I saw the debilitating effect of the way the hospital's treatment of her mum affected her. Together we experienced a 5-year war of attrition with the hospital trust complaint's system. After initially starting with seizures she experienced some appalling attitudes by the medical profession, e.g. the frequent use of the word "pseudo" to describe the seizures, and in one instance a senior A&E operative describing a seizure to junior doctors as "good, but I've seen better".

Joint Perspective. We firmly believe that extreme stress, in our case at the hands of the health system and complaints system, triggered the NEAD attacks. Despite a definitive diagnosis it took some time for local health professionals to understand the illness, and it was we who had to research and provide them with the information. However, the diagnosis has provided us with a positive outlook and this in itself can help to ease the symptoms.

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