CASE REPORT

A Case Report of Munchausen Syndrome by Proxy Presenting as Acquired Symptomatic West Syndrome

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Abstract:

Munchausen Syndrome By Proxy (MSBP) is an extremely complicated diagnosis because of the difficulty in finding the incriminating evidence of its existence and because of the ethical issue it raises for caregivers. Its implications from a medical, psychological and legal point of view raise difficult questions for any professional confronted to it. We present a case of 8 month female infant who was diagnosed to have Hyperinsulinism causing hypoglycemic brain injury and later developing intractable convulsion with head drops, where EEG was suggestive of West Syndrome, was actually a case of Munchausen syndrome by proxy to start with.

Keywords: Munchausen syndrome by proxy, Hyperinsulinism, West Syndrome, Insulinemia.

Introduction:

In 1977 the term Munchausen Syndrome By Proxy (MSBP) was first coined by pediatrician Roy Meadow, an Englishman, when he published a report of a new form of child abuse after the syndrome had been first reported by Asher in 1951 [1]. It describes the deliberate production, or feigning, of physical or psychological symptoms in another person who is under the individual's care. Pediatrician should have a high index of suspicion for this entity since it often produces a diagnostic dilemma in clinical practice. The disorder may be mild where a false medical history is provided or severe where the parent may actually induce the symptom in the child. The fabrication of a pediatric illness is a form of abuse and not merely a mental health disorder, and there is a possibility of an extremely poor prognosis if the child is left in the home.

Case Report:

An 8 month female infant 2nd issue of a non-consanguineous marriage was brought with complaints of one episode of generalized tonic clonic convulsion with history of lithargicity and Global developmental delay, in emergency room. The blood sugar done was low (27mg/dl). Critical samples were sent for investigations such as serum electrolytes, serum cortisol, serum insulin, serum lactate and serum pyruvate and was suggestive of hyperinsulinism (83.6). MRI brain done was suggestive of symemetrical diffusion restriction in bilateral globus pallidus and internal capsule extending into cerebral peduncles probably due to hypoglycemic insult. With initial impression of insulinoma causing hyper-insulinism, she was started on Diazoxide along with Hydrochlorothiazide. Blood sugar levels were well controlled with medical line of management and she had no further episodes of convulsion during hospital stay. Hence she was discharged on Diazoxide and Anti-epileptic drug (Valproate). But after few days of discharging her she again had convulsion and was rushed to hospital. She again had hypoglycemia (25mg/dl) and critical samples were sent again. This time the insulin levels were very high (256.1), which was

thought to be facticious. Again medical history was revised and it was found that the grandfather was a known Diabetic on insulin therapy and the grandmother had frequent quarrels with the mother of the child. Also it was found that every time the infant is kept in house alone with the grandmother the baby had a convulsion. With this history and intense interview sessions with the family members it was found that the insulin was administered to the infant by grandmother causing life threatening complication in the child. The diagnosis of Munchausen syndrome by proxy was confirmed. EEG done, in view of recurrent intractable convulsion and drop attacks and extensor spasms, was grossly abnormal and showed evidence of multifocal epileptiform discharges (Epileptic Encephalopathy) over both hemisphere with burst suppression pattern i.e Hypsarrythmia which is characteristic of symptomatic West syndrome. Injection ACTH was give for a period of 21 days and later was given a course of oral steroid. She remained seizure free during hospital stay and was later discharged on anti-epileptic drug. This is a very unusual case of MSBP.

Discussion:

Children with West syndrome who presented within 1 month from the onset of seizures had better seizure control. Also, children who presented early and had complete seizure control showed good improvement in development. Historically, some previous studies in children with West syndrome, which employed valproate as monotherapy, proved effective in controlling either hypsarrhythmia and/or the epileptic spasms. Besides valproate, zonisamide, topiramate, and nitrazepam (as monotherapy or in combination) showed good responses (ranging from 20 to 35% of treated patients) in West syndrome. More effective, however, has proven the short-term treatment with hormonal therapy (i.e., by using the adrenocorticotropic hormone ACTH), which has been reported to succeed in 60–80% of the infants with West syndrome treated. Therefore, nowadays the first-line treatment of West syndrome and more in general of infantile spasms includes ACTH as well as vigabatrin (the latter being effective especially in infantile spasms in the setting of tuberous sclerosis). Therefore, most recently the treatment of West syndrome with valproate has been overtaken and overlooked.

MSP is a form of child abuse that presents along a continuum from mild to severe. For severe cases, in which the child is clearly being harmed by actions caused or instigated by adults, the appropriate response is to engage the child protection and legal systems. Such "textbook" cases carry a poor prognosis without removal of the child from the caregiver. Detailed management discussions are beyond the scope of this article, but are referenced [1-4]. The expanded concept of MSP provides a more hopeful approach. The Royal College of Pediatrics and Child Health suggest a new nomenclature, i.e., fabricated or induced illness by careers, shifting the focus to child abuse that happens in medical setting [5]. It minimizes harm to the child regardless of the motivation of the perpetrator [6]. Child protection services and legal services may be involved depending on the severity of MSBP. We made use of the familial support system i.e., the mother, so as to safely place back the child with the family. It is concluded that the medical professionals should consider the possibility of MSBP along with their primary differential diagnosis, rather than diagnosis by exclusion.

References

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