Dissociative-like Spells in a Child With Neurofibromatosis (type 1)

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ABSTRACT

Neurofibromatosis, type 1, (NF1) is a common neurocutaneous disorder of childhood. Little is known about the psychiatric aspects of the condition. We present the case of a 10-year-old male with NF1 and complex spells. For two years he had been experiencing self-limited paroxysms of auditory and visual hallucination, assaultiveness, excited undressing, and amnesia. The spells have been refractory to multiple treatments, including antipsychotic medication. The question remains whether this episodic amnestic disorder is comorbid with NF1 or is caused by it.

Neurofibromatosis is a relatively common neurocutaneous disorder that consistently has its onset in childhood. Most common is type 1 (NF1), also known as von Recklinghausen's neurofibromatosis, which accounts for 85 percent of cases and has an incidence of 1 in 3000 persons (1). Penetrance of NF1 is complete in those with the mutation, whether the mutation is sporadic or familial in origin.

Phenotypic expression of the disorder varies greatly, even among family members (2). Clinical features of NF1 are decreased stature, scoliosis, hypertension, cognitive impairment, learning disability, and seizure disorders. Benign and malignant tumors of central nervous system (CNS) and non-CNS origin may occur (3-5). Little is known about the relationship between the disease and psychiatric disorders. Mouridsen and Sorensen (6), in a review of the literature, describe associations between NF1 and autism, learning disability, and attention deficit/hyperactivity disorder (ADHD). In one study, 23 (33%) of 69 adult patients with NF1 had comorbid psychiatric illness: depressive syndromes, anxiety with somatic complaints, and organic

brain syndrome(6). No such studies in pediatric populations have been published. Here, we describe a 10-year old boy with newly diagnosed NF1 and unusual, dissociative-like spells.

CASE REPORT

TP was a 10-year-old, Hispanic boy who had recently been admitted to the neurology service of our children's hospital for workup of several generalized tonic-clonic seizures, occuring earlier that same day, and of abdominal pain. The pediatric psychiatry consultation-liaison service was requested to evaluate the patient's behavioral changes over the past two years. According to the patient's mother, TP had, during that time, episodes of dramatic alterations in his behavior which were inconsistent with his normal personality. These episodes, which sometimes awoke him from sleep, lasted from 10 minutes to several hours and occurred approximately once or twice a week. During these episodes he became aggressive, unheeding of verbal commands from others, and violent toward his younger siblings. His mother said that he often claimed to hear voices of someone or something telling him to hurt his family and to see monsters, from which he would attempt to flee. Sometimes during a spell he undressed and ran into the street. After each episode, and for none of them was there, he said, any recall, he was completely normal (apart from some periods of oppositional behavior).

A seizure had been diagnosed by electroencephalogram (EEG) when he was three. Several different antiepileptics had been prescribed since, but his mother could not recall details about them. At admission he was taking valproate 500 mg twice daily, oxcarbazapine 900 mg twice daily, and risperidone 0.5 mg at bedtime. Nonepileptic seizures had also been documented in the past, according to his mother. No discernable temporal association linked epileptic seizures, nonepileptic seizures, and the amnestic spells described above. Epileptic seizures were reportedly well-controlled on his current medications, but occasional nonepileptic seizures continued to occur.

TP was described as a well-behaved and happy child. ADHD had been diagnosed at a younger age, but the symptoms had remitted by approximately age 8. Conversion disorder had been diagnosed to account for the nonepileptic seizures. On the one hand he was said to be a good student, with no history of disciplinary issues, and on the other to have been in special education classrooms; this apparent discrepancy was not clarified.

No family history of psychiatric or seizure disorders was evident. TP and his mother denied that he had ever suffered physical or sexual abuse.

During the interview, TP was sitting in his bed, playing. He was polite and cooperative. Mood was "happy," affect had normal range. Speech was normal in tone, volume and rate. Short-term and long-term memory were intact, with the exception of amnesia for the spells described above. He showed good concentration and appeared to have normal intellect. Thought processes were logical and sequential. Auditory and visual hallucinations were denied, and no evidence of delusions was apparent. Suicidal and homicidal ideation were absent. He exhibited no insight about the amnestic spells. Outside the spells, his judgment was appropriate.

During the evaluation we observed a typical episode. He slowly fell forward from a seated position and became unresponsive to his mother, the nurse, and those conducting the interview. After several seconds he stood on the bed and hit his mother and baby sister. Over the next few minutes, he threw several items, climbed on furniture, licked the bottom of a shoe, and finally undressed from the waist down and ran into the halls. He was brought back to his bed by hospital staff; then he suddenly calmed, looked down, noticed he was naked, appeared embarrassed, and covered himself with the bedsheet. He denied recollection of the previous few minutes and was completely alert. The baseline level of appropriate affect and behavior returned.

Physical examination was normal for age with the exception of multiple café-au-lait spots on the skin of his lower abdomen, upper right thigh, and back. Freckling in the right inguinal region was also noted. Complete blood count, comprehensive metabolic panel, lipid panel, and urinalysis were normal. There was a slightly elevated serum level of amylase with normal lipase. Valproic acid level was 133 mcg/ml (therapeutic range for seizure control is 50 – 100 mcg/ml). TSH was 4.9 mcg/dl (reference range 0.5 – 4.5 mcg/dl), but when repeated three days later it was 2.7 mcg/dl. The pediatric gastroenterology consultants found no explanation for abdominal pain. An upright radiograph of the abdomen was normal, as was as an abdominal CT-scan. In the images of the CT-scan of the head, ventricles and sulci appeared mildly prominent. Sleep-deprived EEG showed no epileptiform activity.

He was cleared medically and transferred to the inpatient psychiatric unit for further evaluation. A 24-hour, ambulatory EEG was recorded, and during this time a seizure-like episode, resembling a generalized tonic-clonic convulsion, and one of the typical spells described above, took place: the

EEG was normal in both situations. After seven days, he was discharged. Valproate and oxcarbazapine were continued; risperidone was increased to 0.5 mg twice daily to target the oppositional behavior. One month later his mother reported little change in his behavioral symptoms. Brain-MRI was unremarkable.

DISCUSSION

The differential diagnoses for these unusual amnestic spells, we believe, should include seizure disorder, conversion disorder, dissociation, psychosis, malingering, and factitious disorder. Owing to the brief duration and episodic nature of the symptoms, none of the psychotic disorders described in the *Diagnostic and Statistical Manual of Mental Disorders* IV (DSM-IV) seem likely (7). Additionally, complete amnesia for the episodes is not characteristic of psychosis, nor is the fact that they would sometimes awaken him from sleep. Malingering and factitious disorder also seem unlikely, again, because of the sleep disruption, and also because of the complexity of the symptoms.

The relationship between seizures and NF1 has been well-documented. In a study by Kulkantrakorn and Geller (4), the prevalence of seizures in NF1 (4%) was approximately twice that in the general population. Their findings were consistent with previous studies (8,9). Seizures, especially those arising from the temporal lobe, may produce psychiatric symptoms, including psychosis (10). Oner et al. presented the case of an 8-year-old male with psychiatric manifestations similar to those of TP. This patient had temporal lobe epilepsy causing vivid auditory and visual hallucinations, with complete amnesia for the episodes (11). Another case report describes a 30-year-old woman who presented with Fregoli syndrome, and demonstrated the phenomenon of interictal psychosis that can occur with temporal lobe epilepsy (12). Postictal psychosis, although rare, may occur in epileptics, most often after periods of increased seizure frequency; spontaneous resolution takes place over days to weeks (13). In TP's case, however, no seizure activity was recorded on EEG during the amnestic episode, or at any other time in the 24-hour recording. Diagnostic criteria for postictal psychosis were proposed by Logsdail and Toone: (1) the psychosis occurs within one week of a seizure; (2) the psychosis has a minimum length of 24 hours and maximum duration of 3 months; (3) the mental state is characterized by delirium, by delusions and hallucinations with clear sensorium, or by a mixture of these; and (4) the diagnosis excludes patients

with anticonvulsant toxicity, recent head injury, previous interictal psychosis, or minor status epilepticus on EEG (14). In interictal psychosis, symptoms are present for at least six months, hallucinations are present in clear sensorium, and DSM-IV criteria for one of the psychotic disorders are met (15-16). Thus TP's episodes do not fit the description of postictal or interictal psychosis as these terms are currently defined..

Conversion disorder has a prevalence of approximately 1-3% in childhood, although in one English study it was almost 10% (17). According to DSM-IV, in conversion disorder symptoms and deficits in the patient's voluntary motor function suggest a neurologic condition that the patient does not, in fact, have (7). The presentation of conversion disorder in childhood is generally limited by the child's social skills and perceptions of physical illness (18). While there are many forms of conversion disorder, a study of sixteen children in India found nonepileptic seizures to be the most common presenting symptom (19). Nonepileptic seizure can include unresponsive staring, minor motor movements, bizarre behavior, and generalized movements (20). The complexity of the symptoms in the amnestic spells we have described in TP's case is beyond that usually contained by the term "conversion disorder."

We believe that TP's episodes are very possibly dissociative. Dissociative symptoms include amnesia, fugue, depersonalization, derealization, and identity change (20). In a review of the literature, Bowman and Coons explored the overlap of seizures, nonepileptic seizures, and dissociative states. They found that among adult patients with epilepsy, dissociative symptoms are not more prevalent than in the general population. However, adult patients with nonepileptic seizures have a higher prevalence of dissociative symptoms and disorders. In one study of dissociation in nonepileptic seizure, lifetime occurrences were as follows: fugue, 36%; derealization, 56%; depersonalization, 87%; amnesia for periods in childhood, 73%; and amnesia for periods in adulthood (not including the seizures themselves), 82% (20). The amnesia and personality change seen during TP's episodes, and the transient nature of the episodes, are consistent with a dissociative state. On the other hand, the usual causative factor, developmentally, is missing: dissociative disorders are most often associated with childhood trauma or abuse (21-23), which both TP and his mother have adamantly denied in his case. The most we may say, perhaps, is that clinically the episodes resemble dissociative spells. And we may wonder if NF1, in itself, has been the cause.

SOURCE INFORMATION

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