



# Movement and Neurological Disturbance in a Teenage Patient

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## Abstract

A teenage girl came to a pediatrician presenting with a movement disturbance comprised of extension of the right arm, a fixed left arm, body shaking, grimacing, and whimpering. This began happening 2 years ago and had increased in severity the past week. Her previous EEGs had been within normal limits. One previous doctor had thought this a behavioral disorder and prescribed an antidepressant- the parent declined to give this as they did not believe the diagnosis. The patient was formerly a very good student but had since dropped to barely passing classes. She was seen over the course of 2 months between a pediatrics practice and a children's hospital. The differential ranged from psychological, to metabolic, to autoimmune. Several EEGs had come back within normal limits even during movement spells. Finally, an antibody serology provided a diagnosis of autoimmune encephalitis and treatment was started. The patient has since shown great improvements.

## Introduction

Autoimmune encephalitis is a rare diagnosis. This disease occurs when a person's immune system mistakenly labels a cerebral protein as a pathogen [1]. There are no known triggers for this disease and it can occur sporadically [2]. Associations with other autoimmune syndromes and old age have been proposed, but nothing is concrete [3]. It may be hard to diagnose as the symptoms vary greatly and can comprise almost any neurologic sign. The symptoms may also wax and wane as the body heals and attacks itself in cycles [4]. Labs may also be deceptive as they can be as vague as a high white blood cell count and further studies such as antibody studies and an MRI require an index of suspicion related to this autoimmune encephalopathy [5]. As rare as it, this disease is one with great detriment if not caught and treated. It can cause a variety of neurological symptoms that are easy to mistake for psychosis, metabolic issues, and a plethora of other diseases. Mistreated, this disease can progress and cause lasting harm [6]. Brain damage as well as damage to a patient's reputation and education can be consequences of not considering an autoimmune source and treating. In the worst cases, autoimmune encephalopathy can kill a patient if left untreated [1]. For these reasons, it is important that in a patient without other cause of new onset of neurologic symptoms and high white blood cell count, autoimmune sources should be considered [5].

## Case Study

A teenage girl and former honor student from a stable home comes in for a second opinion of a movement disorder that has been ongoing for 2 years and caused her grades to decrease and her friends to become distant. She has a history of thalassemia minor, but no other pertinent history. Family history is also non-contributory. She is oriented to place and time and can follow commands. Her development thus far has been within normal limits. She has yet to start menstruation. There are no known neurological, psychological, or other issues in the family. Vaccines are up to date. Movement episodes include spasms of right arm extending and left fixed and body shaking, grimacing, whimpering, and bladder control loss at times. These occur randomly and throughout the day with no known triggers. They have increased in frequency from a 2 to 3 times a day several weeks ago to 10 to 11 times a day in recent days. Immediately following these episodes, the patient is herself again. There has been no evidence of a post-ictal state. Because of this, she was diagnosed with behavioral Tic disorder and prescribed Seroquel. Her mother was not convinced or the diagnosis and so the patient was never given Seroquel. On physical exam, the patient was oriented and understood prompts. She was in no acute distress, but she did seem emotionally detached- replying to questions with a flattened affect. Her right optic disc appeared pale. Communication seemed impaired-quiet, slow speech, weak voice, articulation poor, and four-word sentences only. There was Dystonia in right and left upper extremity and the ankle and knee reflexes were +1

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bilaterally. The rest of the exam was within normal limits. Orders for ocular screening and a neurological referral were sent. The patient was advised to be admitted to the hospital and return in 2 weeks.

## Investigations

Given the neurological symptoms, an EEG study was conducted with pushbutton instructions for movement episode occurrences. It showed increase activity with movement and back to baseline with no EEG correlate. Next an MRI was obtained, and it also came back within normal limits. The patient was given valproate orally and haloperidol intramuscularly. An ophthalmologist at the hospital cleared her for the optic disk paleness. She was kept for observation overnight and released 2 days later. She returned to the office complaining of worsening symptoms now including hallucinations. A CBC showed increased white blood cells and metamyelocytes. She was advised to return to the hospital for further studies. This time, there was a high index of suspicion for autoimmune encephalitis: Abnormal movements and hallucinations. An antibody study confirmed autoimmune antibodies to an N-type calcium channel.

## Differential Diagnosis

The first diagnosis made was prior to further testing. Due to the presenting symptoms and the lack of EEG findings the diagnosis was behavioral tick disorder. The parents did not believe this diagnosis and did not try the advised treatment. Further studies revealed neurological deficits, so the diagnosis of Wilson's disease was thought of, but the serological studies did not show abnormalities to support this. Finally, after the CBC came back with elevated white blood cells the differential turned to come variety of encephalitis. The antibody studies confirmed autoimmune encephalitis.

## Treatment

Intravenous immunoglobulin was administered in the hospital and prednisone and Divalproex for symptoms were prescribed for long term treatment of 14 days. The pharmacological treatment was completed, and the patient returned to the office without persisting symptoms. The patient was advised that if symptoms ever present again, she should return immediately. Otherwise she should go on about life as usual and we will hope this was a one-time occurrence.

## Discussion

Other cases of autoimmune encephalitis have created a high index of suspicion for those cases presenting in the elderly and in the autoimmune-afflicted population [3]. However due to this a young girl took over 2 years to get diagnosed. She had no other known autoimmune predispositions, and she was so young that psychosis seemed a more likely diagnosis. It is very fortunate that there seem to have been no lasting deficits. In the past, around 6.5% of patients

with newly onset psychosis have been found to have autoimmune encephalitis [6]. This is an easy mistake to make, considering the relative incidence of psychosis and autoimmune encephalopathy [6]. Some of these may self-resolve, but not all. Some can progress and cause long-lasting damage [7,4]. Many cases have been reported to be in combination with focal seizures [8]. Here, there was no EEG activity associated to lead to the correct conclusion. The white blood cell count combined with symptoms was enough in this case to lead to the diagnosis, but it would be all too easy to miss if one were not looking for the right signs. Although this is a relatively rare disease, it is one that must be considered with a broad and unexplained new onset of psychoses [2]. Autoimmune diseases are more common among women of reproductive age; 5 however; the rarity of this specific one can still lead to misdiagnosis, especially since the symptoms can be very misleading. The reversibility here may be due to excellent response to the prescribed medications. Also, it appears the disease had not progressed to cause lasting damage [2].

## Conclusion

This case exemplifies the need for a differential diagnosis on newly onset broad psychosis to include autoimmune encephalitis. It may present with a vast variety of symptoms and can be remitting and relapsing. With the severity of complications with the untreated disease this is a worthwhile one to include on a differential for anyone with similar constellations of symptoms.

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