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Dates: Received: 14 December, 2016; Accepted: 10 February, 2017; Published: 11 February, 2017

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Keywords: Wilson's disease; Tics; Coprolalia; Tourette's syndrome

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Case Report

A Child of Wilson's Disease Presenting with Coprolalia

Abstract

Wilson's disease is a rare autosomal recessive genetic disorder of copper metabolism. Neurological and psychiatric manifestations can be a presenting manifestation of Wilson's disease. We describe a case of a 13 year old child diagnosed with Wilson's disease who presented with coprolalia (complex vocal tics). Tics can be present in Wilson's disease, but they are usually described to be simple motor tics. Vocal tics especially complex vocal tics and that too specifically coprolalia has not been described previously in the literature to the best of our knowledge.

Introduction

Wilson's disease is a rare autosomal recessive genetic disorder of copper metabolism, which is characterised by hepatic and neurological involvement. It was first described by a British neurologist Samuel Alexander Kinnier Wilson in 1912 [1]. The gene which is defective in Wilson's disease is ATP7B which is located on chromosome 13 and encodes an ATPase involved in transport of copper [2]. The neurological and neuropsychiatric signs are the presenting features in 40–50% of patients with Wilson's disease. Psychiatric manifestations can be in the form of personality changes, affective disorders, psychosis, cognitive impairment and others. A review published in 2014 found the most common psychiatric symptoms as depression (30%), incongruous behavior (30%), cognitive impairment (28%) and irritability (22%) [3]. Neurological manifestations include dysarthria, gait disturbance, dystonia, rigidity, tremor, and dysphagia. Less frequent manifestations are chorea and athetosis. Rare neurological presentations include seizures and pyramidal signs [4].

Case Description

A 12 year old male child who was a diagnosed case of Wilson's disease after a neurological presentation earlier with seizure of generalised tonic-clonic semiology and Kayser-Fleischer ring on slit-lamp examination presented to Psychiatry Out-Patient Department with one month history of acute onset in which the child had started hurling abuses in socially inappropriate situations. The parents were very concerned about this behaviour of the child and would frequently feel embarrassed by it. He would have a premonitory urge to hurl the abuses and he would try to resist by keeping his hand over his mouth but

would fail. The child would be embarrassed by this behaviour and had significant distress due to this. The child would be aware of the phenomenon. Coprolalia (complex vocal tics with obscene or socially inappropriate content) was established. There were no motor tics. There were no significant findings on a detailed clinical neurological examination. The child did not have any other movement disorder. At the time of presentation, there were no clinical features suggestive of hepatic involvement. The liver function tests were normal. Neuroimaging (Magnetic Resonance Imaging) did not reveal any significant abnormality. Serum ceruloplasmin and 24-hour urinary copper were within normal limits. Genetic testing was not performed due to affordability issues. Child had a score of "4" based on Ferenci Scoring system which establishes the diagnosis of Wilson's disease [5]. He was receiving Penicillamine and Zinc.

He presented to our outpatient department. Child was started on antipsychotic in the form of oral Risperidone 1 mg per day on which he improved over a period of 3–4 weeks.

Discussion

Tics are repetitive stereotyped movements or sounds that are sudden, rapid and nonrhythmic and preceded by premonitory sensation or urge. Tics may be motor or vocal; simple or complex. Complex vocal tics consist of well formed words. A specific kind of complex vocal tic is coprolalia which consists of production of obscene words which are socially inappropriate. Coprolalia has been found to be commonly present in Tourette's syndrome where it is associated with multiple motor tics as well. Tics usually have an onset in prepubertal period commonly in ages 4 to 6 years. New onset presentation during teenage is uncommon and presentation in adulthood is rare.

An atypical age of onset raises suspicion for the presence of a specific etiology. In the present case, child presented with new onset tics at the age of 13 years. The tics were complex vocal tics specifically coprolalia. There were no motor tics. The child had already been diagnosed with Wilson's disease. In the index case, neuroimaging did not reveal any abnormality. A study by Kim et al. in 2006 found many children (including some with neurological symptoms) with Wilson's disease having normal MR imaging findings [6]. Serum ceruloplasmin was also within normal limits when he presented to our outpatient department. Serum ceruloplasmin may decrease after initiation of treatment with Penicillamine [7]. Tics can be present in Wilson's disease but they are usually described to be simple motor tics. Vocal tics especially complex vocal tics and that too specifically coprolalia has not been described previously in the literature to the best of our knowledge. A recent review of 20 case series and cohort studies published in 2014 [3] did not report vocal tics. This case highlights the varied presentation of Wilson's disease and also highlights that new onset tics at atypical age should raise suspicion of some other underlying etiology. We managed the case with the usual management of tics that is with low dose antipsychotic. The child had improved with Risperidone 1mg/day. Antipsychotics work in management of tics of varied etiology. Eddy et al. [8], reviewed the treatment strategies for tics and found Risperidone to be efficacious in management of tics. However such cases need to be monitored longitudinally to observe the course and progression of such symptoms. Neuroleptics should be used with caution as there is a risk of worsening of dyskinesias and other extrapyramidal symptoms [9]. It is difficult to comment whether the complex vocal tics in the child were a result of neuropsychiatric manifestation of Wilson's disease or it had developed independently. However the atypical age of onset and already diagnosed status of Wilson's disease makes Wilson's disease the most likely

explanation. The case further illustrates the need to investigate a child for Wilson's disease in a variety of neuropsychiatric presentations.

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