GILLES DE LA TOURETTE'S SYNDROME:

A COMMONLY MISDIAGNOSED NEUROBEHAVIORAL DISORDER

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Abstract: *We report two cases of Gilles de la Tourette syndrome Case 1, thrity year old male, presented with complains of episodes of finger drumming and titubation. Upon further enquiring into the history, the patient recalls history of multiple tics occurring daily since childhood. There was history of obsessive compulsive bhaviour and palilalia. Case 2 is that of a ten year old boy who also presented with multiple motor tics. The parents ggave history suggestive of obsessive compulsive behavior, attention deficit hyperactive disorder and coprolalia. In both case the routine haematological and biochemical investigations were within normal limits. CT scan head and EEG were normal. Based on the clinical presentation a diagnosis of Tourettes syndrome was made and treated with antpsychotic. Both the patients had significant improvement within 1 month of therapy.*

INTRODUCTION

Tourette syndrome (TS) is a neuropsychiatric disorder with onset usually in the first two decades of life characterized by a combination of multiple motor tics and at least one phonic tic. Phonic tics may be simple sounds (simple phonic tics such as grunting or sniffing) or whole words (complex phonic tics). In the latter case they are called complex phonic tics whose most characteristic example is coprolalia (searing or emission of obscenities). Tics are often associated with behavriorl abnormalities such as hyperactivity and attention deficit disorder, obsessive compulsive disorder and others as was seen in both of our cases.

CASE REPORTS

Case 1: A 30 year old male, presented with episodes of involuntary movement of the fingers of the right hand in the form of repeated tapping and turning of the head. Upon further enquiring in to the history, the patient recalls similar involuntary movements, which were identified as tics, occurring since childhood. He first initially noticed the tics when he was 6 years old and had complains of frequent eye blinking . Later he developed tic in the feet and hands. These occurred daily and would cause significant embarrassment. He used to cover up by putting his hands in the pocket and start pacing. He faced difficulty in learning, academics and work performance due to lack of concentration. He also had complex vocal tics and used to repeat ones own utterances particularly the last word (palilalia). There as history suggestive of obsessive compulsive behavior in the form of repeated hand washing due to fear of contamination. Both motor and phonic tics used to exacerbate during times of stress such asexaminatons and interviews, so much so that in one instance he was sent out of the examination hall, suspecting that he was cheating in the exam. He was also refused many jobs and was fired twice from his job owning to his problem. He as born following a fulterm normal delivery and had normal developmental milestones. No family history of any psychiatric or neurological illness. On examination his higher mental functions were intact. His mood was congruent with his affect. There was no abnormality in the thought process. There was no cranial nerve involvement. No sings of Motor, sensory and cerebellar involvement. He had a normal gait. Haematological and biochemical investigations were unremarkable. Ct scan head and EEG were normal. Based on the classical presentation a clinical diagnosis of Tourette;s syndrome was made and the patient was started on antipsychotic medication Resperidone (1mg twice daily). He responded significantly with disappearance of tics within 1 month of treatment. The quality of life has also significantly improved and patient in now performing well academically and proressionally.

Case2.: A ten year lod boy who was brought by the parents with complains of requent eye blinking, head jerking and shoulder shrugging. The parents recall, they first noticed the problem when he was 7 years old. It started with eye blinking and later the lateral head truning and shoulder shrugging. These motor tice used to occur daily. The child was also very fidgety and would get distracted very easily. His teachers complained about his fidgidity and that he lacked concerntraion during his teaching sessions. His academic performance was poor. He was also very impulsive with a low threshold for engaging into fights with his friends. There was history suggestive of obsessive compulsive behavour, felt that his hands are dirty and contaminated to the extent that he suded to wash them 20-30 times a day His parents also complained that their son had started using abusive language,. swearing words for no reason to everyone including his parents because of which he ass punished may a times but was still unable to control himself. All this caused a lot of frustration and embarrassment and now he had very few friends. He as born following a ful term normal delivery. No history suggestive of any perinatal insults and had normal developmental milestones. He has two elder brothers and one younger sister, none of whom exhibit any fetures of aneurobehavioural disorder. On examination his higher mentalfunctions and cranial nerves were intact. No sings of motor, sensory cerebeelar involvement and had normal gait. Upon investigations he had normal haematological and biochemical exams. CT scan head and EEG were normal. A diagnosis of tourette syndrome was made and he was started on haloperidol. He showed significant improvement with 6 weeks with decreased frequency of motor and phonic tics but still had some features of attention deficit disorder. His academic performance has improved and is now getting good grades in school.

DISCUSSION

Tourette syndroms (TS) named after George gilles de la tourette who in 1885 coned this syndrome of persistent but changing tics and coprolalia beginning in childhood. It is a neuropsychiatric disorder with onset usually in the first two decades of life characterized by a combination of multiple motor tics and at least one phonic tic. This syndrome is often misdiagnosed as a psychiatric illness2 A. family

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history is sometimes obtained, and partial expression of the trait may occur in siblings or offsprings of patients. In most families with chronic tic disorders, there is an autosomal dominant mode inhertiaance with variable penetrance that is gender related. Although the disorder has been reported in association with chromosomal abnormalities, in vast majority of cases the chromosomes are normal3 boys are affected much more commonly than girls. On the basis of the etiology, ST is classified into two catergories. idiopathic and symptomiatic symptomatic TS can occur after head inury encephalitis stroke, rheumatic fever, and drug intake (Methylphenidate, neuroleptics, opioids, antiepileptics), Motor tics commonly affect the face and may consist of repetitive snfiffing, winking blinking, elevation of eyelids eye closure, pursing of lips or fcial twitching, patients eventually develop several different motor and phonic tics, the latter frequently taking form of grunts, barks, hisses, sighs, throat clearing, coughing, phonic tics may be simple sounds (simple phonic tics such as grunting or sniffing) or whole words (complex phonic tics) In the latter case they are called complexphonic tics whose most characteristic example is coprolalia (swearing or emission of obscentities) It must be emphasized that although possibly the most characteristic symptom of Tourette syndrome4 coprolalia is not mandatory for the diagnosis of this codition. The pathophysiology of coprolalia and other complex phonic tics remains ill defined. The tics may change in location, severity, complexity and character with time are worsened by emotional stress and can be suppressed voluntarily for short periods of time. Tics are often associated with behavioral abnormalities such as hyperactivity and attention deficit disorder, obsessive compulsive disorder and others. These co morbid conditions offer interference in learning, academic and work performance. it has a life long course with remissions and exacerbations. Many patients have non specific electroencephalographic abnormalities of no diagnostic significance . Treatemnt responses is seen with high potency antipsychotic durgs including pimozide, haloperidol and respiridone.

There have been many cases reported in the Indian literatute. A recent study by Anand et al5 showed the clinical profile of the cases reported in India is similar to those reported from the western counties with the exception of coprolalia and ADHD, the incidence of which is higher in the cases reported in India. The pathophysiology is obscure and no structural pathology has been reconginzed. A dopaminergic excess has been suggested by the clinical observation that the tics may respond to treatement with dopamine blocking drugs.

In another report by Duggal eta al6, they have studied the Bereitschafts potential, movement related cognitive potential indicative of motor preparation, in three patients with Tourette syndrome and have implicated quasi volitional nature of tics and that they are not totally involuntary, Desai et al7 described the use of metoclopramide, a potent dopamine antagonist, in patients with Tourette syndrome.

Kumar s8 reported Neurofibromatosis type1 manifesting with Tourette syndrome, a rare presentation, and pointed towards the pathological involvement of caudate and lentiform nuclei in the genesis of tics in TS,

CONCLUSION

Gilles de la Tourette's syndrome is often misdiagnosed as a case of psychiatric disorder, as was the scenario in our patient. These patients are subjected to unnecessary, expansive investigations and treatment before the corret diagnosis is made, Depression developed in our patients, due to the social embarrassment cause by the tics, which could have easily been avoided with the early recognition of this syndrome.

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DRUG PROFILE

Rifaximin

Pharmacokinetics: Rifaximin, a semi-synthetic, nonsystemic antibiotic. Rifaximin can be administered with or without food. Systemic absorption of Rifaximin was low in both the fasting state and when administered within 30 minutes of a high fat breakfast. There is no evidence of accumulation of Rifaximin following repeated administration for 3 days (9 doses). Peak plasma Rifaximin concentrations after 3 and 9 consecutive doses ranged from 0.81 to 3.4 ng/mL on Day 1 and 7.83 + 4.94 ng-h/mL on Day 3. Rifaximin is suitable for treating systemic bacterial infections because less than 0.4% of the drug is absorbed after oral administration. Animal studies have that 80% to 90% of orally administrated Rifaximin is concentrated in gut with less than 92% in the liver and kidney, and less than 0.0 1% in others tissues. In adults with infectious diarrhiea treated with Rifaximin 800 mg daily for three days at Rifaximin is excreted primarily in the feces. After oral administration of 400 mg. ***Indications:*** Escherichia coli infection of the guity to the main indication Rifaximin to healthy volunteers approximately 97% of the dose was recovered in feces, almost entirely as unchanged drug, and 0.32% was recovered in the urine. ***Special Populations***: ***Geriatric*** : The pharmacokinetics of Rifaximin in patient> 65 years of age has not been studied. *Pediatric:* The pharmacokinetics of Rifaximin has not been studied in pediatric patients of any age. *Renal Insufficiency:* The pharmacokinetic of Rifaximin in patients with impaired renal function hasbeen studied. *Hepatic Insufficiency*: Rifaximin plasma concentrations of 13.5 ug/mL. were detected in hepatic encephalopathy administrated Rifaximin 800 mg three times daily for 7 days. Drug-Drug Interactjns, In an in vitro hepatocyte induction model Rifaximin was shown to induce cytochromic P450 3A4 (CYP3A4), an isoenzyme which Rifampin is known to induce. Two clinical drug interaction studies have been conducted using midazolam, Rifaximin has not shown to significantly affect intestinal of hepatic CY3A4 activity. The oral contraceptive showed study that the phannacokinetic of single dose of ethinyl estradiol and gestimate were not altered by Rifaximin ***Dosage and Administration:*** Rcifax tablets can be administered orally with or without food. For travelers diarrhea, the recommended dose is one 200 mg tablet taken three times a day for 3 days. ***Adverse Effects***: Common side effects include nausea, flatulence, constipation, vomiting, pyrexia, headache, LyIi/abdominalsain, rectal tenesmus, defecation urgency. ***Warning:*** The drugs is in effective in cases of travelers, diarrhea due to Campylobactojejunis; Should be discontinued if diarrhea persists; antibiotic therapy should be considered. Pseudomembranous colitis has been reported with use of antibiotics due to the overgrowth of nonsusceptible organisms; should superinfection occur during therapy, appropriate measures should be taken.