CASE PRESENTATION

Tardive Dystonia: The Significance of Early Recognition and Management

Abdulrahman Al Tahan, MRCP*
Hassan Al Maleh, MD, FAMS Psych*

ABSTRACT

Tardive dystonia is a recently well described complication of antipsychotic medications. Compared with the long and better known tardive dyskinesia, it is more disabling and difficult to manage. We report the development of tardive dystonia in a young Saudi male with schizophrenic disorder. The favourable outcome after abrupt discontinuation of antipsychotic medication is documented. An early sign of repetitive forced jaw opening is reported. Literature on tardive dystonia is reviewed with special emphasis on the significance of early recognition and management in improving the prognosis.

Tardive dyskinesia (TDK), was described in late 1950s, as a complication of the long term use of antipsychotic medications[1]. Tardive dystonia (TDT), which is a later onset and persistent dystonia occuring in the same setting, was noted much later[2], and it tool until 1982 to be recognised as a subtype of TDK of special significance[3]. However, since then, awareness of this entity increased and reports accumulated describing its different features. In comparison with TDK it was noted to be less common but more serious and difficult to treat, making it one of the more feared complications of antipsychotic drugs, causing considerable disability and hindering the social adjustment of affected patients[4-6]. This situation enhances the role of prevention and early management especially in relation to early recognition and prompt discontinuation of antipsychotic medications, as was stressed by several workers[6-8]. To illustrate these points, we will describe a well-documented case of young man with schizophrenic disorder who developed TDT six years after being started on triflouperazine. An early sign, antedating the development of the full clinical picture is noted.

THE CASE

A 28 year old man was followed up since the age of 22 at our hospital with the diagnosis of probable schizophrenic disorder. His symptoms included persecutory delusions, threatening hallucinations, incoherent speech, inappropriate smile, lack of volition, and deterioration in social functioning and school performance. Previously he was on trifluoperazine upto 45mg and benzhexol 6mg daily in the previous six years. These medication were continued after reducing the doses. The patient was regular in his follow-up appointments, and was complaint with medications. He was able to hold a part-time job, and

* The division of Neurology and Psychiatry
King Khalid University Hospital
King Saud University
Riyadh, Saudi Arabia

his behaviour was generally acceptable for the next four uyears while on trifluoperazine 10mg per day, and benzhexol 4mg per day most of the time. At the age of 26, the patient's relatives noticed that he started to have frequent episodes of jaw opening sustained for few minutes which was confirmed by the patient, who described having intermittent abnormal feeling of jaw stiffness. Trials of increasing and reducing the dose of benzhexol during the following six months gave no apparent response. The patient got married and stopped taking his medications. Within days he started to have mild forced turning of neck to the left side. When assessed, he was found to have repetitive, intermittent, forceful and painful deviation of the neck to the left side, which improve on supporting the neck with both hands, and become worse on walking. He also had upward contraction of both eyebrows, repetitive grimacing, intermittent pouting and difficulty in swallowing. He was videotaped and scored by the treating team, using the abnormal involuntary movemnets scale (AIMS)[9], estimating the severity as 22/42.

Apart from these abnormal movements, he was neurologically well and was psychologically in reasonable remission. His past history included normal milestones with no incident of head or neck injury. None of his family had any neurological disease, especially movement disorder.

The patient had a long list of normal investigations which included: liver and renal functions assessment, serum copper, ceruloplasmine and head CT scan. The final diagnosis was TDT related to the prolonged use of trifluoperazine. In the following few months, the patient was regularly followed while on diazepam 15-25 mg daily only. A gradual improvement of his movements was noted and by the end of six months, his AIMS score was 16/42. However as his wife left him, he relapsed into psychotic episode which was treated with sulpirid 800 mg a day. This was reduced to 200 mg b.i.d as he was controlled, in combination with diazepam 15 mg and benzhexol 8 mg per day. His movements showed further improvement with an AIMS score of 12/42. Eyebrows movement disappeared and his torticollis, pouting, grimacing, and jaw stiffness became mild. This was maintained over the next six months of follow-up.

DISCUSSION

Being a relatively newly recognised entity, TDT may still be passed unnoticed, labelled as TDK which is the well known long term complication of antipsychotic medications. However in a recent study, TDT prevalence in chronic psychiatric patients was found to be 2% (10), which incrase the importance of identifying TDT especially in view of the implications regarding management and prognosis[6,11]. The diagnosis of TDT in this patient was based on the criteria formulated by Burk et al[3], which includes: (i) the presence of chronic dystonia, (ii) its temporal association with the use of antipsychotic medications, (iii) exclusion of known causes of secondary dystonia, and (iv) negative family history of dystonia.

Dystonia may be easily confused with hysteria, and proper assessment of the clinical setting and possible gain are important[12]. In our case, the patient's social life and adjustment were seriously affected by the development of TDT, while his psychiatric illness may reasonably controlled. Acute dystonic reaction can be easily excluded on account of the following points: first, oculogyric crisis do not occur in TDT; second, the onset of acute reactions occur within 24 to 48 hours of exposure to the antidopaminergic drugs, while TDT usually occurs after long period of exposure. However contrary to conventional belief, TDT may occur earlier, but never reported within the first 48 hours[6]; thirdly, TDT persists for long periods, rarely remitting, while the acute dystonic reactions disappear on discontinuing the offending agents[6]. TDT should be differentiated from other causes of chronic dystonia. Of special importance in this region is Wilson disease which is considered locally to be relatively more common here than elsewhere. An impression echoed earlier by Bahemuka et al[13]. Other entities to consider include Parkinson disease, Hallervorden-Spatz disease, cerebral lipidosis, Huntingtonchorea, and intracranial structural lesion. Primary dystonia should also be considered especially in the presence of family history. Clinically TDT differs by its early involvement of the face or neck, being less generalised, and commonly associated with oral dyskinesia. It is also less severe and rarely leading to a chronic bedridden state, as is the course commonly in primary dystonia[3]. This patient exhibits some of the associations noted to occur more in TDT than with TDK including the prolonged use of antipsychotic drugs with short interruptions[14], and the young age and male sex[3,5,14], though the last two points were not substantiated by Kang et al[6]. However, other reported associations including depression, poor response to antipsychotics, previous use of electro-convulsive therapy, and the presence of organicity were absent[15-17].

Neuropathology of TDK in general is thought to result from an increased responsiveness or supersensitivity of dopamine receptor sites, caused by long-term neuroleptic blockade[18]. More recently, experimental data suggested a role for GABA ergic system[19].

Early recognition of TDT is important in view of the strong suggestion that early dicontinuation of antipsychotic medications is a favourable factor in remission[6]. Some workers even considered it as the major variable in determining reversibility[7]. Reports of early signs, described restricrted vermicular movements of tongue[20], and eye blinking preceding by two years the full picture of TDT[21]. In a series of 11 patients with TDK[14], frequent eye blinking was reported in seven patients, twitching of the face and extremities in two, unspecified neck movements in one and lip smacking in another. In our patient "intermittent jaw opening" appeared six months before the clear features of TDT were established, which seem to have been accelerated by the prompt discontinuation of his medications[4]. However, his symptoms went on to improve persistently after cessation of trifluoperazine. Whether this improvement is related also to the use of diazepam will be difficult to ascertain. However, diazepam was only occasionally of benefit in treating TDT[6], making the role of the prompt, discontinuation of antipsychotic drug, the significant factor. When the need for reinstituting antipsychotic medications arises, a different and relatively safer drug should be used[4].

Treatment of TDT is generally disappointing as can be guessed from the long list of tried agents[22-28]. Burk et al, found that of all agents used in their study, dopamine depleting drugs (tetrabenzine and reserpine), and anticholingergics were the most effective, though only rarely did they cause remission or satisfactory control of symptoms[3].

This account of TDT stresses the importance of prevention. Kang et al, found that 22% of their TDT patients were prescribed antipsychotic medication for inappropriate indication, such as anxiety and nausea[16]. These medications should only be given after deliberate consideration and only if no alternative is available.

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