Olanzapine Treatment in Choreoathetosis due to Bilateral Striopallidodentate Calcinosis

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ABSTRACT:
Olanzapine treatment in choreoathetosis due to bilateral striopallidodentate calcinosis

Bilateral Idiopathic Striopallidodentate Calcinosis (BISPDCs) refers to idiopathic calcification of the basal ganglia and secondary striato-dentate calcinosis. Its definition comprises numerous metabolic abnormalities including disturbances of calcium and phosphorous metabolism. In this article, a case with BISPDCs secondary to hypoparathyroidism who presented with generalized choreoathetosis has been reported. A daily dose of 2.5 mg of olanzapine has been initiated and increased gradually up to doses 10 mg/daily. In six weeks time, the patient was almost free from choreoathetosis. Thus, olanzapine may be a good choice at the symptomatic treatment of choreoathetosis due to BISPDCs.

Key words: striopallidodentate calcinosis, olanzapine, choreo-athetosis

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INTRODUCTION

Bilateral idiopathic striopallido-dentate calcinosis (BISPDCs) refers idiopathic calcification of the basal ganglia (1-3), and secondary striato-dentate calcinosis. Its definition comprises numerous metabolic abnormalities including disturbances of calcium and phosphorous metabolism (4). The most common neurological symptoms of BISPDC include parkinsonism, chorea, dystonia, ataxia, seizures, mental deterioration and pyramidal findings (5). Movement disorders accounted for 55% of the total symptomatic patients' (6). In hypoparathyroidism and pseudohypo-parathyroidism the diminution in ionized serum calcium induces not only tetany and seizures but sometimes choreoathetosis as well (%50), and also in some instances there are signs of a cereballar lesion (7). The pathophysiology of this condition remains unknown and results of treatment are often unsatisfactory (8). Some authors suggest that the pathogenesis of cognitive and motor changes in this disease are based on a dysfunction of cortico basal connections and their interhemispheric relations (9).

Atypical antipsychotics have greater activity in blocking serotonin-2A (5-HT_{2A}) receptors than dopamine-2 (D_2) which mitigates extrapyramidal symptoms. They also block D_2 receptors long enough to cause an antipsychotic action. The increased sensitivity to dopamine as a result of treatment with conventional antipsychotics drugs may not occur with atypical antipsychotics (10). The effect of olanzapine in improvement of chorea also suggests that olanzapine has a dopaminergic D_2 receptors blocking action (11). Olanzapine treatment in choreoathetosis with tardive dyskinesia and Huntington's disease have been reported before (12-14). However its effect in choreoathetosis due to BSPDC has not

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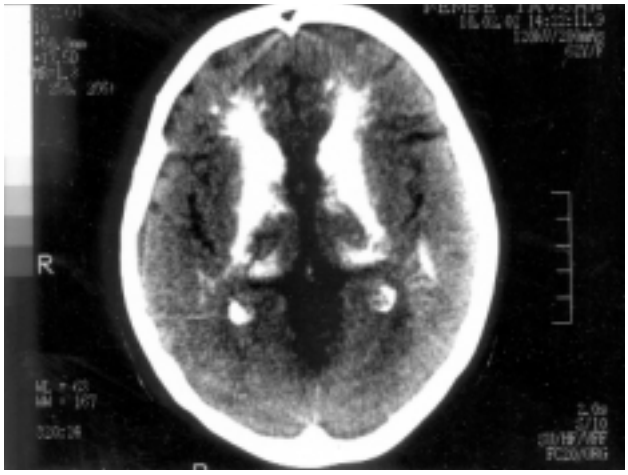
been published before. Here we report a case with bilateral striato-pallido-dentate calcification secondary to hypoparathyroidism who presented with generalized choreoathetosis and who prominently improved after olanzapine treatment.



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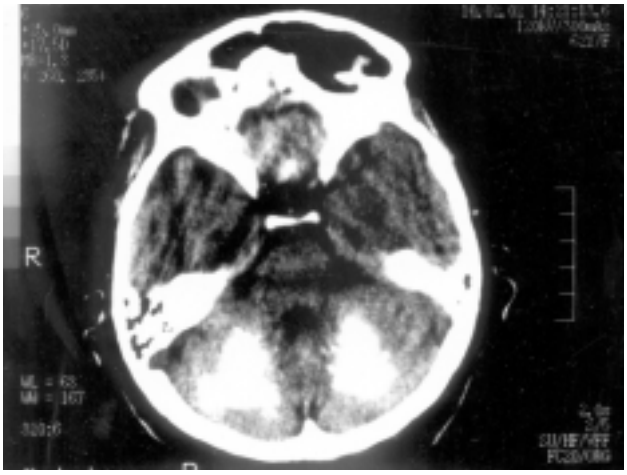

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Figure 1. CT scan images

CASE REPORT

A 66-year-old woman referred to our hospital with the complaint of involuntary movements at her limbs and face. She had a thyroidectomy operation 37 year ago. She have been suffering from hypocalcaemia secondary to iatrogenic hypoparathyroidism that first occurred a few days after her thyroidectomy operation. There were many episodes of hypocalcaemia and hypocalcemic tetany in her past medical history. These attacks had been treated with calcium infusions and then put on calcium supplementation therapy. She also had thyroid hormone replacement and orally calcium therapy for 37 years. She received the diagnosis of depression and used various antidepressant drugs, but she have not used any antipsycothic drugs. There was no history of familial chorea.

On admission she was suffering from involuntary movements at all four limbs and orobuccal muscles. She informed that these involuntary movements were first started at her mouth six months ago and than diffused to her extremities and progressed worse during last one month. The choreiform orobuccal and limb movements were observed on her neurological examination. Abnormal Involuntary Movement Scale (AIMS) score was 2 at her mouth, 4 at her right upper and lower extremities and 3 at her left upper and lower extremities. These involuntary movements were obvious at her right side and did not show any change with motor action. Other findings in the neurological examination were unremarkable.

Serum calcium and parathormone levels were low, and serum phosphorous level was high. Her CT scan revealed symmetrical calcifications in the basal ganglia, thalamic nuclei and cerebellar-cerebral white matter (Figure 1). We started 2.5 mgr daily olanzapine up to doses 10 mgr/day. One week later buccal choreiform movements disappeared while limb movements persisted with reduced severity (AIMS scores were as follows: right side leg: 3, arm: 3, left leg: 2, arm: 2). Six weeks later the patient was almost free from choreoathetosis (AIMS score was 2 at her right leg) and she was able to do her daily activities. We did not determine any side effect during this therapy and the patient was discharged with 10mg/day oral dose of olanzapine therapy. The case was followed about six months and after one month from discharging she was almost free of involuntary movements. Thus the dose of olanzapine was reduced 2.5mg. While she was on 7.5mg daily dose of olanzapine, she did not suffer from any side effect, and continued the therapy at the same dose. The patient did not show up on regular controls after six months.

CONCLUSION

Basal ganglia calcification with hypoparathyroidism initially pointed before and occurrence of basal ganglia calcification in hypoparathyroidism is well known (15). Calcification is most commonly seen at globus pallidus but other structures can be involved (2). The defective iron transport and free radical production may cause tissue damage and calcification (16). About half of patients with basal ganglia calcification have neurological features. Headache, vertigo, movements disorders, stoke-like events, seizure and syncope are reported most common clinical manifestations. Extrapyramidal movement disorders occur in 56% of the patients of the movement disorders; parkinsonism accounted for 57%, chorea 9%, dystonia %8, athetosis 5% and orofacial dyskinesia 3 % (6,15,16).

There is no specific treatment to limit calcium accumulation (16). Symptomatic treatment of BISPDC related parkinsonism often responds to levodopa (16) but atypical antipsychotics for the therapy of these patients may be a good choice because these patients are sensitive to extrapyramidal system side effects during clinical antidopaminergic medication (16). Atypical antipsychotics have greater activity in blocking 5-HT_{2A} receptors than D_2 which mitigates extrapyramidal symptoms and atypical antipsychotics block D_2 receptors long enough to cause an antipsychotics action (10). Olanzapine is a serotonin-dopamin receptor antagonist similar to clozapine and there are reports about olanzapine treatment in choreoathetosis with tardive dyskinesia and Huntington's disease (12-14) but its effect for choreoathetosis due to BSPDC has not been published before. Some authors suggest that the pathogenesis of cognitive and motor changes in this disease is based on a dysfunction of corticobasal connections and their interhemispheric relations (9,11). Atypical antipsychotics have several important clinical benefits, including lesser extrapyramidal side effects and dysporia, less impaired cognition, and a lower risk of tardive dyskinesia than typical antipsychotics (17). Pharmacologically, different atypical antipsychotics differ; these differences covert into differences in their side effect profiles (17).

Olanzapine, is one of the novel atypical with significantly fewer treatment-emergent extrapyramidal symptoms and less akathisia associated with traditional antipsychotics (18). Compared with traditional agents, olanzapine shows only a few adverse events such as dry mouth, sedation, and increase in appetite and compared with risperidone, olanzapine causes greater increases in weight gain and body mass index but less hyperprolactinemia (18). Transient, non-dose-dependent, asymptomatic elevations in liver enzymes have also been noted in olanzapine-treated patients.

Depression stands for a complex of disturbed feelings like dispair, hopelessness, a sense of worthlessness, and thoughts of self-harm, which are associated with, decreased energy and libido (7). Our patient had the diagnosis of chronic depression by a psychiatrist, and she was still suffering from unhappiness, sense of worthlessness and decreased energy. Her Hamilton depression scale score was 14. Tetrabenazine, which is a dopamine depletor, was another alternative drug for this patient but we did not prefer it, because tetrabenazine could aggravate her depressive symptoms.

As a result we emphasize, olanzapine is a reasonable alternative therapy for the symptomatic treatment of choreoathetosis due to Bilateral Strio-Pallido-Dentate Calcinosis.

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