Deep Brain Stimulation in Multiple System Atrophy: A Rare Case Report with Positive Clinical Response to STN-DBS

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Abstract

Introduction:Multiple system atrophy (MSA) is a progressive neurodegenerative disorder characterized by autonomic dysfunction, parkinsonism, and cerebellar ataxia. Current treatment options are limited and fail to significantly alter the disease course. While deep brain stimulation (DBS) is widely accepted in the management of Parkinson's disease (PD), its efficacy in MSA remains controversial, with only sporadic reports of clinical benefit.
Method: A 53-year-old female patient presented with a 5-year history of rigidity and bradykinesia, initially responsive to Levodopa. She subsequently developed severe dyskinesias, dysphagia, dysarthria, and postural instability. Initially diagnosed with PD, the diagnosis was later revised due to clinical deterioration and MRI findings of pontine atrophy with the "hot cross bun sign," suggestive of MSA. In 2023, she underwent bilateral subthalamic nucleus (STN) DBS implantation using stereotactic techniques and microelectrode recording. The procedure was uneventful, and device activation occurred 14 days postoperatively. Following activation, complete control of dyskinesias was achieved, with immediate symptom recurrence upon DBS deactivation. Marked improvement in bradykinesia and gait was observed, with subsequent symptom stabilization. Rigidity and dyskinesia control have remained satisfactory to date, contingent upon continuous stimulation. Interestingly, shortly after surgery and before stimulation onset, the patient experienced a transient worsening of bulbar symptoms (dysphagia and dysarthria), with gradual recovery to preoperative baseline within two months. Over the long term, these symptoms have progressed slowly, though less severely than typically expected in MSA. To date, the patient has maintained sustained improvement in motor symptoms, representing a rare positive outcome in DBS for MSA.
Discussion: Although MSA is traditionally regarded as poorly responsive to DBS, this case demonstrates that STN stimulation may yield significant motor benefits in selected patients, particularly when diagnostic overlap with PD exists. The favorable response highlights the importance of comprehensive, multidisciplinary evaluation in selecting candidates for DBS in complex neurodegenerative disorders.
Conclusions: While DBS is not disease-modifying in MSA, it may serve as an alternative therapeutic strategy in carefully selected cases with predominant parkinsonian features, contributing to improved functional status and quality of life.

References

1. Di Luca DG, Ramirez-Gomez C, Germann J, Santyr B, Boutet A, Milosevic L, et al. Deep brain stimulation of the globus pallidus internus and externus in multiple system atrophy. Mov Disord. 2023;38(11):2121–5. doi:10.1002/mds.29573. PMID:37544011
2. Badihian N, Jackson LM, Klassen BT, Hassan A, Low PA, Singer W, Coon EA. The effects of deep brain stimulation in patients with multiple system atrophy. J Parkinsons Dis. 2022;12(8):2595–600. doi:10.3233/JPD-223504. PMID:36442207; PMCID:PMC10077954
3. Ullman M, Lewis SJG, Dewey RB, Guttman M, Stoessl AJ. Deep brain stimulation response in pathologically confirmed cases of multiple system atrophy. Parkinsonism Relat Disord. 2012;18(1):86–8. doi:10.1016/j.parkreldis.2011.09.007
4. Lezcano E, Gómez-Esteban JC, Zarranz JJ, Alcaraz R, Atarés B, Bilbao G, et al. Parkinson's disease-like presentation of multiple system atrophy with poor response to STN stimulation: a clinicopathological case report. Mov Disord. 2004;19(8):973–7. doi:10.1002/mds.20108. PMID:15300669
5. Tarsy D, Apetauerova D, Ryan P, Norregaard T. Adverse effects of subthalamic nucleus DBS in a patient with multiple system atrophy. Neurology. 2003 Jul 22;61(2):247–9. doi:10.1212/01.wnl.0000073986.74883.36.
6. Moretti DV. Available and future treatments for atypical parkinsonism: a systematic review. CNS Neurosci Ther. 2019;25(2):159–74. doi:10.1111/cns.13068.