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Globus Pallidus Internal Deep-Brain Stimulation in a Patient with Neuroacanthocytosis with Drug-Induced Parkinsonism

 Paresh Doshi^a Nikhil Chamankar^b
^aDepartment of Neurosurgery and Stereotactic and Functional
Neurosurgery, Jaslok Hospital and Research Centre, Mumbai,
India; ^bDepartment of Neurosurgery, Jaslok Hospital and
Research Centre, Mumbai, India

Contracted red cells with a number of irregularly spaced, thorny surface projections, often with terminal bulbs, are defined as acanthocytes. Acanthocytosis is generally related to 3 hereditary neurological disorders: abetalipoproteinemia, chorea-acanthocytosis (ChAc), and Hallervorden-Spatz disease. Here, we present the case of a 77-year-old female who had been suffering from orofacial and neck dyskinesias secondary to neuroacanthocytosis for 3 years. Two years later, she started having difficulty in walking and showed body stiffness. On examination, she was found to have orofacial dyskinesias, dyskinesias of the neck, and tremors in the right upper limb. She had slurred speech, walked independently but with difficulty, and dragged her right foot. Her off-phase Unified Parkinson Disease Rating Scale (UPDRS) motor score was 36. Her laboratory findings were suggestive of neuroacanthocytosis.

The orofacial dyskinesias, for which she was treated with tetrabenazine, started 3 years back. After 2 years of treatment, she developed asymmetric, Parkinsonian symptoms, as described above. At the time of admission, she was on 37.5 mg tetrabenazine, 3 mg trihexyphenidyl, and bendroflumethiazide 75 mg/day. However, none of these medications could control her symptoms. She had also been given a trial of levodopa, but it worsened her orofacial symptoms. Her medical management was getting increasingly difficult, as medical treatment for PD would worsen the orofacial dyskinesia and vice versa. We offered her left globus pallidus internal (GPi) deep brain stimulation, as it was thought to be beneficial for both pathologies. After thorough evaluation, the patient underwent left GPi deep-brain stimulation. Following the surgery, her dyskinesias were completely resolved, and there were minimal signs of residual PD. The patient continued to do well, and her symptoms were well controlled for up to 5 years of follow-up. During this time, there was a gradual increase in her Parkinsonian symptoms, requiring increased stimulation parameters (from 3.0 V initially to 5.3 V) and the introduction of levodopa. At the last follow-up, she was on 300 mg levodopa, 37.5 mg tetrabenazine, and 1.5 mg pramipexole.

Table 1. Stimulation parameters at the time of IPG replacement (5 years after surgery)

Channel	Contacts	Voltage, V	Frequency, Hz	Pulse width, μ s	Limits, V
1	C+1–2–3–	5.3	130	90	0–5

She required an implanted pulse generator change after 5 years. At that time, her settings were as described in Table 1. Alteration in the globus pallidus/substantia nigra pars reticulata complex due to imbalance between direct and indirect motor pathways led to hyperkinetic orolingual movements [1]. Drug-induced Parkinsonism (DIP) is the second most common cause of PD and is known to occur in patients on tetrabenazine [1]. Though DIP is bilateral, it can be asymmetric and unilateral [1]. The diagnosis can be made by the chronology of the appearance of the symptoms and the use of dopamine transporter imaging. In our patient, PD started 2 years after tetrabenazine, and dopamine transporter imaging also confirmed the diagnosis of DIP. She was highly dependent on tetrabenazine and hence had to be offered some solution that could control both symptomatology. It is known that GPi stimulation is effective in dyskinesias secondary to neuroacanthocytosis and controlling symptoms of PD [2, 3]. Miquel et al. [4] showed that there was long-term improvement in gait disturbances and orofacial movements following GPi stimulation in patients with chorea-acanthocytosis. This is the first case report of GPi stimulation showing long-term sustained benefit in controlling the symptoms of chorea-acanthocytosis and DIP.

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