Benefit of bilateral pallidotomy in the treatment of generalized dystonia

Case report

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This 29-year-old man with cerebral palsy complicated by generalized dystonia was treated by simultaneous bilateral posteroventral pallidotomy. Postoperatively, there was slow, but steady, improvement in the patient’s dystonia and disability. However, the improvement in abnormal movements was only prominent for cervical dystonia and oromandibular dyskinesia. The patient’s Burke-Fahn-Marsden dystonia scores were 51 preoperatively and 37, 33.5 and 33.5, at 3, 6, and 12 months postoperatively, respectively, demonstrating a maximum improvement of 34%. These results suggest that pallidotomy can be an alternative therapy for those patients suffering from intractable generalized dystonia.

KEY WORDS • cerebral palsy • dystonia • pallidotomy
cerebral atrophy, but there was no lesion noted in the basal ganglia. We chose to perform a simultaneous bilateral PVP to treat the patient’s dystonia. The Burke-Fahn-Marsden Evaluation Scale for dystonia was used for pre- and postoperative objective assessment.

Before the operation, the patient underwent brain MR imaging (1.5-tesla imaging unit; Siemens Magnetom, Erlangen, Germany). The detailed anatomy of the basal ganglia was visualized using inversion recovery sequences obtained with a long acquisition time to provide high-resolution images. The relationship between the posteroventral portion of the inner segment of the globus pallidus (GPi) and the anterior and posterior commissures of the third ventricle were calculated, and the predicted lesion volume was defined for each side. A stereotactic frame (Leksell G stereotactic frame; Elekta Instruments, Stockholm, Sweden) was secured to the patient’s head after application of a local anesthetic agent and he was transferred to the MR imager, where the coordinates of the anterior and posterior commissures were established.

Operation. The tentative target was chosen at the center of the posteroventral portion of the patient’s GPi. It was 2 mm anterior to the midcommissural points and 21 mm lateral and 5 mm inferior to the intercommissural line. With the patient receiving local anesthetic, a small burr hole was made in the frontal skull at a point 2.5 cm away from the midline and near the coronal suture. A thermocoagulation bipolar probe (2 mm in diameter with a 4-mm uninsulated separation between the anode and cathode [Elekta Instruments]) was placed at the center of the posteroventral portion of the GPi through the burr hole. Electrical stimulations were made to check the response first in the internal capsule (8 Hz, 300 μsec, up to 10 amp) and second in the optic tract (80 Hz, 200 μsec, up to 10 amp). If there was no sign of muscular contraction of contralateral limbs or eye flushing, a reversible lesion was made by thermocoagulating the target at 60°C for 60 seconds. When the preservation of good motor function of the contralateral extremities was confirmed, permanent lesions (80°C for 80 seconds) were simultaneously made.

Postoperative Evaluation and Course. Postoperatively, an MR image confirmed accurate placement of the thermocoagulated lesions in the bilateral posteroventral portions of the GPi (Fig. 1). There was no immediate complication, including cerebral hemorrhage. The patient was discharged from the hospital 2 days after surgery. Clinically, there was a dramatic reduction in the muscular hypertonus of the patient’s extremities and neck and his dystonia began to improve 3 months later, followed by additional slow but steady improvement. The maximum improvement in the dystonia, approximately 34% over baseline, was noted at 6 months postsurgery (Table 1). The improvement in the dystonia was most notably observed as a nearly complete remission of cervical dystonia and oromandibular dyskinesia. However, there was no significant improvement noted in the patient’s arms, trunk, or legs. His speech became much clearer, but was still a little difficult to understand. His masticating and swallowing movements were better but choking still occurred occasionally. These clinical improvements have persisted for 12 months following surgery. There has been no adverse effect of the surgery noted on the patient’s motor and visual functions during the entire observation period.

Discussion

Stereotactic surgery on the posteroventral portion of the GPi to control parkinsonian symptoms was initiated by Lars Leksell in the 1950s. However, further development of this approach was halted because of the introduction of medical treatment with levodopa. In the early 1990s, Laitinen and colleagues began to reevaluate the therapeutic effect of pallidotomy in patients with Parkinson’s disease (PD). Since then, PVP has been proven to be effective in ameliorating response fluctuations, hypokinesias, rigidity, and levodopa-induced dyskinesia in patients with PD. Moreover, PVP has also been found to be effective in relieving muscular pain and dystonia. Because of its safety and lack of significant side effects, PVP can be considered as an alternative strategy for treating dystonia.

Posteroventral pallidotomy, as primary therapy for dystonia, has been investigated during the past few years. Iacono, et al. reported sustained relief of all dystonic symptoms and abnormal movements in a young male patient with dystonia muscularum deformans who underwent simultaneous bilateral PVP. Lozano and colleagues also used the surgery to treat a young boy with severe generalized dystonia. In that case, a 79% improvement in both axial and limb dystonia was noted 3 months after the operation. The present study showed a similar result for bilateral simultaneous PVP, leading to a 34% improvement in abnormal movements in a patient with intractable generalized dystonia. Furthermore, the study supports PVP as an alternative treatment for generalized dystonia.
A similar result was reported by Iacono, et al.,6 that bilateral PVP produced a generalized improvement in patients with PD.7,8,10,12 However, recent studies reversing akinesia and eliminating dyskinesia simultaneously that PVP directly reduces GPi activity, thereby resulting in motor function in idiopathic dystonia is different from that of PD. Hashimoto and Yanagisawa1 showed that overactivity of the premotor cortices, as a result of underactivity of the basal ganglia, was found both at rest and during movement in idiopathic dystonia. Microelectrode recording during PVP in the study by Lozano and coworkers6 showed that the firing rate of GPi neurons in a patient with generalized dystonia was lower than that in patients with PD and in normal primates. Furthermore, surgical intervention with PVP, which disrupted the altered neuronal activity of the GPi, could remarkably improve motor function in patients with dystonia. Despite the lack of microelectrode recording data in the present study, we observed a remarkable improvement in the abnormal movement in this patient following PVP. The finding may imply that neuronal activity in the GPi plays an important role in the mechanism of dystonia and that a reduction in the altered neuronal activity in the GPi can improve motor function in patients with dystonia.

Conclusions

This study shows that bilateral PVP is an effective treatment for generalized dystonia, although the improvement in motor function in our patient was mostly limited to the craniocervical region. Based on our observations in this patient, we suggest that bilateral PVP warrants further investigation as a new treatment modality for generalized dystonia.

References


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TABLE 1
Burke-Fahn-Marsden dystonia scores in a patient who underwent bilateral posteroventral pallidotomy for generalized dystonia

<table>
<thead>
<tr>
<th>Region</th>
<th>Maximum Possible</th>
<th>Preop Baseline</th>
<th>Mos Postop 3</th>
<th>Mos Postop 6</th>
<th>Mos Postop 12</th>
</tr>
</thead>
<tbody>
<tr>
<td>eyes</td>
<td>8</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>mouth</td>
<td>8</td>
<td>4</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>speech/swallow</td>
<td>16</td>
<td>9</td>
<td>6</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>neck</td>
<td>8</td>
<td>8</td>
<td>2</td>
<td>0.5</td>
<td>0.5</td>
</tr>
<tr>
<td>rt arm</td>
<td>16</td>
<td>12</td>
<td>9</td>
<td>9</td>
<td>9</td>
</tr>
<tr>
<td>lt arm</td>
<td>16</td>
<td>8</td>
<td>8</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>trunk</td>
<td>16</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>rt leg</td>
<td>16</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>lt leg</td>
<td>16</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>total</td>
<td>120</td>
<td>51</td>
<td>37</td>
<td>33.5</td>
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