

Nonketotic Hyperglycemic Hemiballism Treated with Stereotactic Pallidotomy

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Hemiballism is an unusual involuntary movement. Diabetes mellitus (DM) induced hemiballism is rare and only 23 patients with the disorder have been reported in the literature. We report two patients with nonketotic hyperglycemic hemiballism. They were refractory to medicine for four and six months respectively even though their serum glucose levels were well controlled. Therefore, they underwent stereotactic pallidotomy for the treatment of these hemiballisms. Magnetic resonance imaging (MRI) revealed a characteristic hyperintensity lesion on T1-weighted images on the contralateral side of the basal ganglia. These are the first two published cases of hemiballism treated with stereotactic pallidotomy. Both patients had satisfactory outcomes and returned to normal daily life. (*Mid Taiwan J Med* 2003;8:53-8)

Key words

hemiballism, non-ketotic hyperglycemic, stereotactic pallidotomy

INTRODUCTION

Hemiballism has an incidence rate in the order of 1 in 500,000 of the general population [1] and it occurred in about 0.7% of patients with movement disorders in a series by Dewey [2] at the Baylor Movement Clinic. Hemiballism is a hyperkinetic disorder, characterized by regular, wide-amplitude, vigorous movements of limbs, with involuntary muscular construction of proximal limbs and head [3]. The movements often occur simultaneously on one side of the body. The most common feature of hemiballism is destructive lesion [4-6], usually vascular in origin and most commonly found on the contralateral side of the subthalamic nucleus [1]. Other locations include the basal ganglia [7], lenticular nucleus [8-10], caudate nucleus [11] or subcortical white mater [12]. Metabolic disorders [13] such as thyrotoxicosis, systemic lupus

erythematosis, uremia, and dysfunction of glucose metabolism rank are known to cause this hyperkinetic disorder. Nonketotic hyperglycemic hemiballism has rarely been reported [14-20], however it has stimulated more interest recently due to the characteristic hyperintense lesions on T1-weighted images (T1WI) on the contralateral side of the basal ganglia [21,22]. Treatment of hemiballism is usually with medication but not all drugs are effective. Stereotactic surgery is another method to treat hemiballism that, until now, has not yet been applied to nonketotic hyperglycemic hamiballism. We present two patients with nonketotic hyperglycemic hemiballism who were successfully treated with stereotactic pallidotomy.

CASE REPORT

Case 1: A 70-year-old male with long-term DM suffered from involuntary movement of right limbs for six months after an episode of hyperglycemic hyperosmolar non-ketotic acidosis (HHNK). His serum glucose had not been well controlled (ranged: 185 mg/dL to 334 mg/dL). He took Haloperidol 5 mg tid for two months to

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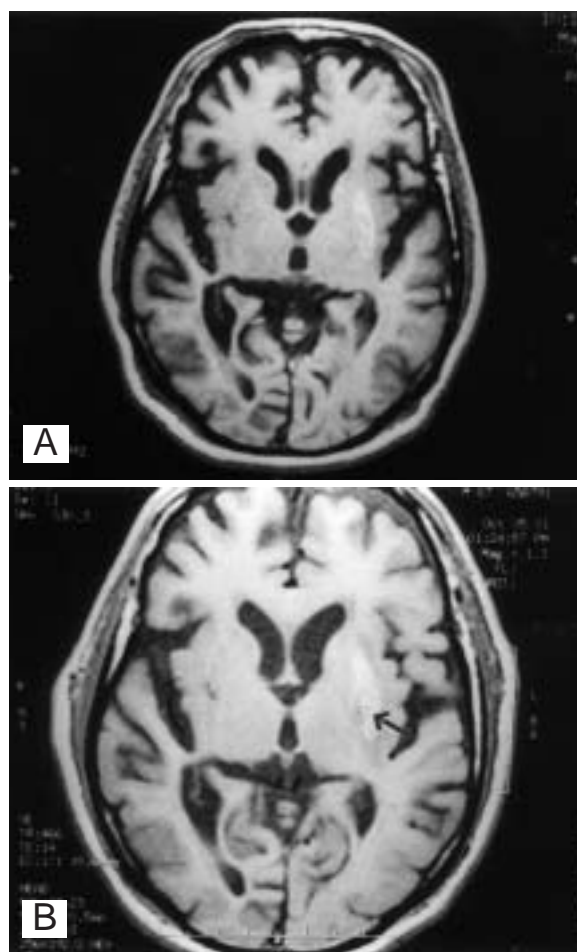


Fig. 1. A: Pre-operative MRI of case 1 shows a hyperintensity lesion in the left corpus striatum on T1WI; B: The post-operative MRI on T1WI shows stereotactically created lesions (arrow head) in the left-lateral globus pallidus by radiofrequency electrocoagulation.

control the hemiballism but it was not effective. The restless involuntary movement of his right limbs made it difficult for him to eat, sit, and walk. The movement ceased during sleep. The Hemiballism/hemichorea outcome rating score (HORS) was graded as 3/5, (normal = grade 0/5). MRI showed a typical hyperintensity lesion on left basal ganglia. He underwent stereotactic pallidotomy under local anesthesia. The hemiballism disappeared immediately after the pallidotomy with 80°C and 60 sec radiofrequency thermocoagulation. He was discharged uneventfully one week later. The preoperative and postoperative MRI are shown in Figs. 1A, 1B.

Case 2: A 76-year-old female suffered from involuntary movement of her right limbs for one

month. She had a history of non-insulin dependent diabetes and developed right hemiballism with symptoms of uncontrollable head movements. A few days before admission, proximal, repetitive, stereotyped flinging movements of the right arm and leg developed. Grimacing of the right side of the face, blinking of the right eyelid and slow rotating movements of the right foot were also found. These involuntary movements were so violent that they made sitting, eating, speaking, and falling asleep difficult; however, they ceased during sleep.

The severity was graded as 4/5 using HORS [5]. Serum glucose level was 263 mg/dL, and was controlled with 8 units of regular insulin and 16 units of NPH. She was given 5 mg Haloperidol Tid for involuntary movements without effect. MRI showed a typical hyperintense lesion at the left globus pallidus. Valproate, Chlorpromazine, and Diazepam were ineffective. Therefore she underwent stereotactic pallidotomy (Fisher's stereotactic frame) with radiofrequency thermocoagulation (Radonic). The target lesions were set on the globus pallidus for 6 points with 2-mm bipolar tip. The coagulation temperature and duration were 80°C for 60 seconds for each point. Following stereotactic pallidotomy, 2 mg Dexamethasone qid daily for three days was prescribed to decrease the brain edema.

Hemiballism subsided on the first postoperative day. Only intermittent slow movement of her right foot remained. At that time the HORS decreased to grade 1 and totally disappeared two weeks later. The pre-operative and post-operative MRI are shown in Figs. 2A, 2B. She was discharged after an uneventful course. The HORS improved to grade 0. She is now able to walk and eat by herself, and leads a normal daily life.

DISCUSSION

Cases of nonketotic hyperglycemic hemiballism are rare in the literature [14-20]. Most of the patients were more than 60 years old (mean age: 65.1 years) and predominately women (M/F = 10/15). Limb involvement was more common than face or trunk. The prognosis of

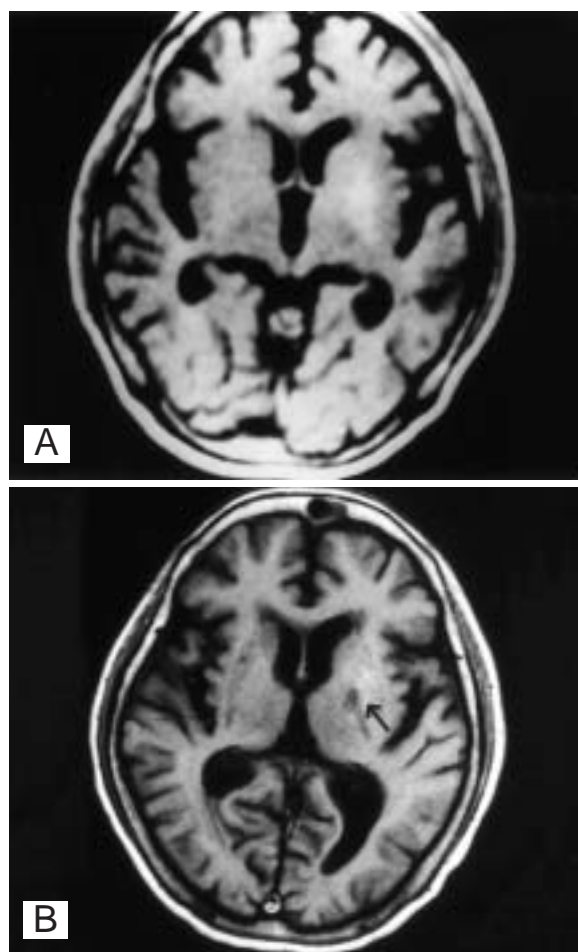


Fig. 2. A: Preoperative MRI of case 2 on T1WI; B: postoperative MRI on T1WI. Operation lesions are marked with arrow head.

hemiballism induced by hyperglycemia is as good as hemiballism induced by infarction or other causes. Usually involuntary movement subsides within a few days after lowering serum glucose levels with intravenous insulin and hydration. In a series by Lin and Chang [20], the symptoms of three patients with hemiballism caused by HHNK, disappeared within three days after hyperglycemia control. Some patients need continuous anti-dopaminergic drugs to control the hemiballism. In a series by Vidakovic et al [6], one patient was treated with Haloperidol and hemiballism disappeared within two weeks. In a series by Yahikozawa et al [22], three patients received Haloperidol, Chlorpromazine, and Sulpiride, respectively, all these medications

achieved good outcomes. Hemiballism may be recurrent when serum glucose levels elevate once again [11,19].

In our patients, the involuntary movements were so severe that daily life quality was poor even after four to six months of medication. Both of them suffered from deep depression because these involuntary movements disturbed their walking, sitting, eating, and other daily activities. Therefore, stereotactic pallidotomy was applied for them.

In a recent report by Krauss and Mundinger [5], 13 (93%) out of 14 patients improved or had no residual symptoms after stereotactic thalamotomy. Two out of 14 patients in their series received stereotactic thalamotomy combined with pallidotomy. Long-term follow-up results (mean follow-up period, 11 years), revealed that 12 out of 13 patients have had persistent improvement in hemiballism; seven patients (54%) were free of symptoms and five patients (39%) had minor residual involuntary movements. However, none of the patients in the above surgical reports had hyperglycemic induced hemiballism. Our two cases are the first nonketotic hyperglycemic hemiballism patients treated with stereotactic pallidotomy.

Current models of functional organization of the basal ganglia remain inadequate to explain the various effects of functional stereotactic surgery on movement disorders. Some researchers have targeted the anterior portions of the pallidum [23], the medial pallidum [24,25], and the lateral pallidum [23-26]. All of these target lesions were effective in ablating hemiballism. Pallidotomy for treatment of hemiballism is intended to interrupt pallidothalamic output [27]. On the other hand, stereotactic thalamotomy reduces excessive thalamocortical drive [5]. The optimal method of treating hemiballism is still a controversial issue. The side effects of medial pallidotomy and thalamotomy are mild hemiparesis and dystonia, which frequently subside or disappear during the follow-up period.

MRI image findings of hyperglycemic hemiballism present unique pictures in the

contralateral striatum [22]. They are characterized by a persistent increase in intensity on T1WI and a slight decrease in intensity on T2WI. The striatal lesions are presumed to have developed following mild ischemia in the territory of the lateral striatal branches of the middle cerebral artery [22]. These characteristic MRI findings of the striatum facilitate our location for pallidotomy.

Hyperglycemic hemiballism occurs more frequently in older women. Estrogen can decrease the dopamine (DA) function of the nigrostriatal system and subsequently increase the density of DA receptors [28,29]. Concentrations of estrogen decrease in women after menopause, which contributes to the development of supersensitivity in the striatal DA receptor [30]. DA hypersensitivity in postmenopausal women is presumed to be factor for developing this type of hyperkinesias [20]. It may also explain the fact that hyperglycemic hemiballism occurs more frequently in women older than 50 years. The supersensitive striatal lesion of the DA receptor may be destroyed by pallidotomy with thermocoagulation. This was the theory behind the treatment chosen for nonketotic hyperglycemic hemiballism in our patients. Using the same theoretic base, we can explain the successful treatment of patients with anti-dopaminergic drugs such as Haloperidol [2,31], and Reserpin [32]. Although the actual mechanism of pallidotomy for treating hemiballism needs further evaluation, pallidotomy was effective in treating nonketotic hyperglycemic hemiballism in our patients.

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以立體定位蒼白球燒灼術治療因非酮類高血糖引起之 半邊舞蹈症

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半邊舞蹈症是不尋常之不自主運動。因糖尿病病人導致之半邊舞蹈症是很罕見，目前文獻上只有廿三例報告。我們報告兩例因非酮類高血糖引起之舞蹈症，他們分別經過四個月及六個月藥物治療，雖然其血糖已獲得控制但仍無效，才考慮用手術治療。所以他們經過立體定位手術之蒼白球燒灼手術來治療半邊舞蹈症，在磁振造影上其病灶有特殊表徵，就是在T1影像上之對側蒼白球位置呈現高信號，此種信號有利於立體定位燒灼。我們所報告之二例，是目前文獻上僅有二例用立體定位蒼白球燒灼來治療半邊舞蹈症之病例。他們皆獲得良好之預後且恢復正常生活。（中台灣醫誌 2003;8:53-8）

關鍵詞

立體定位蒼白球燒灼術，半邊舞蹈症，非酮類高血糖症

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