CASE REPORT

Hemiballism with leg predominance caused by contralateral subthalamic haemorrhage

Kazuyuki Noda,¹ Nobutaka Hattori,² Yasuyuki Okuma³¹Department of Neurology, Juntendo University Shizuoka Hospital, Izunokuni, Japan²Department of Neurology, Juntendo University School of Medicine, Bunkyo-ku, Japan³Juntendo University Shizuoka Hospital, Izunokuni, Japan

Correspondence to

Dr Kazuyuki Noda,
k-noda@juntendo.ac.jp

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SUMMARY

Hemiballism is a rare movement disorder characterised by high-amplitude movements of the limbs on one side of the body. Stroke of the contralateral basal ganglia, especially the subthalamic nucleus (STN) is the most common aetiology of acute development of hemiballism. Recently, the pathophysiology of hemiballism has been associated with abnormal firing patterns in the globus pallidus interna, with intermittent firing bursts followed by pauses, during which movements occur. An 87-year-old woman presented with a 5-day history of hemiballism predominantly in her leg. On the basis of her brain MRI findings, she was diagnosed as having vascular hemiballism caused by haemorrhage in the contralateral STN. Treatment with risperidone led to the clinical resolution of her condition. We discuss the distribution of the patient's ballistic movements on the basis of the somatotopic organisation of the STN.

BACKGROUND

Hemiballism is characterised by violent and involuntary flinging movements of one side of the body, and stroke is the most common cause of this movement disorder. Historically, hemiballism has been classically considered as one of the most localisable symptoms in neurology, and pathognomonic of a lesion of the subthalamic nucleus (STN).¹ However, several current case series have challenged the paradigm of hemiballism resulting from contralateral STN lesions.² Stroke localised to the STN was found by neuroimaging in only 4 of 27 patients reported by Ristic *et al*,³ 4 of 27 reported by Chung *et al*,⁴ 4 of 22 reported by Vidaković *et al*,⁵ 4 of 21 reported by Dewey and Jankovic,⁶ 4 of 11 in the Lausanne stroke registry⁷ and 2 of 15 reported by Postuma and Lang.^{8,9} It is considered that reduced or impaired STN excitation of the globus pallidus interna (GPi) leads to abnormal firing rates and patterns. The random or disorganised firing patterns from GPi to the motor nuclei of the thalamus cause fluctuating patterns of inhibition and disinhibition of the motor cortex, which are believed to cause hemiballism.² Interestingly, there are few case reports in which the association between the site of hemiballism and somatotopy in the STN is discussed. We report the unusual case of a patient presenting with hemiballism, predominantly in one leg, caused by a small focal haemorrhage.

CASE PRESENTATION

An 87-year-old right-handed woman with a history of hypertension was admitted 5 days after the

abrupt onset of involuntary movements of her right leg. On neurological examination, there were rapid, uncontrollable and flinging movements predominantly in the right leg, and minimally in the right arm. Involuntary movements were not seen in the axial musculature or face (video 1). The ballistic movements were accentuated by any attempt to move, decreased at rest and disappeared during sleep. There were no abnormal eye movements on ocular examination. No neurological deficits were found except for the movements.

INVESTIGATIONS

Results of laboratory tests, including complete blood cell counts, serum electrolyte levels and liver function tests, were normal. On the day of admission, brain MRI with T2-weighted imaging and fluid-attenuated inversion recovery (FLAIR) imaging showed a high-signal-intensity area surrounded by oedema in the left STN. The maximum diameter of the haemorrhage itself was 5 mm, and the maximum diameter including the surrounding oedema was 15 mm (figure 1A, B). Gradient-echo (T2*) MRI showed hypointensity in the left STN (figure 1C). The brain MRI with FLAIR imaging showed a focal and discrete high signal intensity in the left STN (figure 1D). We diagnosed the patient as having vascular hemiballism caused by a minute haemorrhage in a certain part of the STN.

TREATMENT

The patient had not been treated with antiplatelets or anticoagulants. After admission, she was treated with 1 mg of risperidone once a day with blood pressure monitoring.

Video 1 Our patient, an 87-year-old woman, is shown. Prior to the initiation of the treatment, she demonstrates ballistic and choreatic movements predominantly in the right leg, at rest and during action.

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Figure 1 MRI of the patient 5 days postevent. Axial T2-weighted (A), fluid-attenuated inversion recovery (FLAIR) (B), gradient-echo (T2*) (C) and coronal FLAIR (D) MRIs show a minute acute haemorrhage in a certain part of the left subthalamic nucleus. One scale expresses 10 mm.

OUTCOME AND FOLLOW-UP

The abnormal movements resolved over the next 2 weeks following administration of 1 mg of risperidone once a day. The patient was transferred to a nearby hospital for further rehabilitation.

DISCUSSION

The clinical feature of our patient was abrupt-onset hemiballism caused by a contralateral STN lesion. The hemiballism of this patient was predominant in one leg. In previous reports, ischaemic lesions were responsible in the majority of cases, whereas haemorrhagic lesions were responsible in less than 20% of reported cases.¹⁰ There have been several reported cases of patients with small isolated haemorrhagic lesions in STN.^{10–17} Moreover, hemiballism predominantly in one leg caused by an isolated haemorrhage of the STN is very rare. Ohnishi¹² and Maruyama *et al*¹³ reported cases of patients with ballism confined to the contralateral leg, so-called monoballism. In the case reported by Ohnishi, a 62-year-old man developed monoballism in his right lower limb. His MRIs showed a single haemorrhage in the caudal portion of the contralateral STN without other associated lesions. In the case reported by Maruyama *et al*, a 62-year-old man also developed monoballism in his right lower limb. His MRIs showed a dumbbell-shaped haemorrhage extending from the contralateral STN to the dorsomedial thalamus. In addition to the haemorrhage, there was accompanying

oedema extending to the globus pallidus, which resulted in a much bigger affected area. The distribution of the hemiballism in our patient was closely similar to that in the reported cases of patients presenting with monoballism, although the arm was minimally involved. In the patients reported by Ohnishi, Maruyama *et al* and in our case, the causative lesion for contralateral ballism was considered to be the one in the STN, but the size on MRIs in each of these cases slightly varied.

If highly ordered somatotopic representations in the STN exist, it will be helpful to elucidate how lesions restricted within this nucleus result in impaired movements of a single body part. Human autopsied cases with partial damage to the STN showed that hemiballism exhibits a somatotopic pattern, so that damage to the rostral, middle and caudal portions of the STN affects the face, arm and leg, respectively.^{18–19} Although no correlation has yet been revealed between the site of the STN lesions and the somatotopic specificity of hemiballism, dyskinesias experimentally produced by STN lesioning showed the following characteristics: (1) dyskinesias appear predominantly in the lower and upper limbs, but rarely in the orofacial area; (2) lower limb dyskinesias are more marked than upper limb dyskinesias; and (3) upper limb dyskinesias usually occur in association with lower limb dyskinesias.^{20–24} Interestingly, Nambu *et al*²⁵ demonstrated a clear somatotopy in the primate STN. The primary motor cortex (M1) projects to the lateral half of the STN, whereas neuronal projections from the supplementary motor area (SMA) are

predominantly distributed within the medial half of the STN. The orofacial-associated, upper limb-associated, and lower limb-associated regions of the M1 project to the lateral to medial parts of the lateral half of the STN, whereas those of the SMA project to the medial to lateral parts of the medial half of the STN. Therefore, two sets of somatotopic representations, which are mirror images of each other, are represented in the lateral and medial parts of the STN. When the mediolateral central zones of the STN are damaged, the lower limb-associated regions of the M1 and SMA domains are affected by simultaneously formed lesions. On the other hand, large lesions that infringe on a considerable part or on the entire mediolateral extent of the STN are perhaps required to destroy the upper limb-associated and/or orofacial-associated regions of both the M1 and SMA domains. It is conceivable that a given body part may be affected when STN lesions encompass the homotopical zones of both the M1 and SMA domains simultaneously. In the human STN, cells associated with leg movements are located medially and centrally, and cells associated with arm movements are distributed more diffusely.²⁶ We speculate the mechanism underlying the leg predominance of hemiballism in our patient as follows. First, the haemorrhage itself was minute and localised in the leg-associated part of the contralateral STN. Second, the accompanying oedema finally resulted in a much bigger affected area. Partial dysfunction of the portion of the STN relevant to face and arm movements inhibited ballism, which should have appeared in the face and arm. Further studies are required to clarify the correlation between the distribution of hemiballism and somatotopy in the STN.

Learning points

- ▶ Hemiballism is a rare movement disorder characterised by unilateral flinging movements of the limbs.
- ▶ Hemiballism is most commonly caused by stroke. Causative lesions are located in the putamen, caudate nucleus, cerebral cortex, thalamus and subthalamic nucleus.
- ▶ Hemiballism predominantly in one leg is rare and highlights the complexity of the pathophysiology of hemiballism.
- ▶ Precise diagnosis by MRI is crucial for the early treatment and favourable prognosis.

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Competing interests None.

Patient consent Obtained.

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