

Case Reports

Ipsilateral Hemichorea-hemiballism in a Case of Postoperative Stroke

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Abstract

Background: Ipsilateral hemiballismus refers to the rare occurrence of hemiballism developing on the same side of a brain lesion.

Case report: We describe a rare case of postoperative ipsilateral hemiballism in a patient who underwent pituitary adenoma resection and experienced a right internal cerebral artery territory infarct. We review the literature on hemichorea hemiballismus (HCHB) and explore various mechanisms for its occurrence.

Discussion: Only three cases of ipsilateral hemiballism have been described, and the exact pathophysiology remains unknown. A dominant left hemisphere with corpus callosal connections to the right basal ganglia is the most probable explanation for this unusual event.

Keywords: Hemiballismus, hemichorea, stroke, pituitary adenoma, surgery

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Introduction

Hemichorea-hemiballism (HCHB) is a spectrum of involuntary, continuous, nonpatterned movement involving one side of the body. It usually results from a lesion in the contralateral subthalamic nucleus (STN) and adjacent structures.¹ Ipsilateral hemiballismus is very rare; only three cases have been reported. We encountered this rare entity in a patient who underwent pituitary adenoma surgery complicated by a right internal carotid artery (ICA) territory infarct. In this report, we review the HCHB literature and propose a possible explanation for this condition.

Case report

A 23-year-old female presented with headache for 2 years and drooping of the right eyelid and visual blurring for 2 months. She had associated amenorrhea and coarsening of facial features. On examination, she was confused, irritable, had no light perception in the right eye, and 6/36 vision in the left eye. Visual field examination revealed a left temporal field defect, and the right eye could not be evaluated due to poor vision. Fundus examination revealed right optic

atrophy. Brain computed tomography (CT) and magnetic resonance imaging (MRI) revealed a sellar and suprasellar mass with right cavernous sinus extension, suggestive of large pituitary adenoma (Figure 1).

She underwent a right pterional craniotomy and gross total tumor decompression. Intraoperatively, the mass had soft to firm consistency. The lesion was predominantly extradural in some areas where it had breached into the subarachnoid space. The right ICA was encased by the tumor and was also decompressed. There was no obvious direct injury to the ICA during surgery. Postoperatively, the patient was drowsy but moved both her upper and lower limbs in response to painful stimuli. She suffered sensory deterioration over the next few hours and developed left-sided hemiplegia. A repeat CT scan showed hypodensities in the right ICA territory indicating a massive right ICA infarct, and decompressive craniotomy was immediately performed. On the second postoperative day, she developed rapid right-sided involuntary movements of both the upper and lower limbs. These consisted of flexion and extension, rotation, crossing, and were predominantly distal (chorea). This was admixed with severe, violent, arrhythmic, and large amplitude movements of the proximal limb with

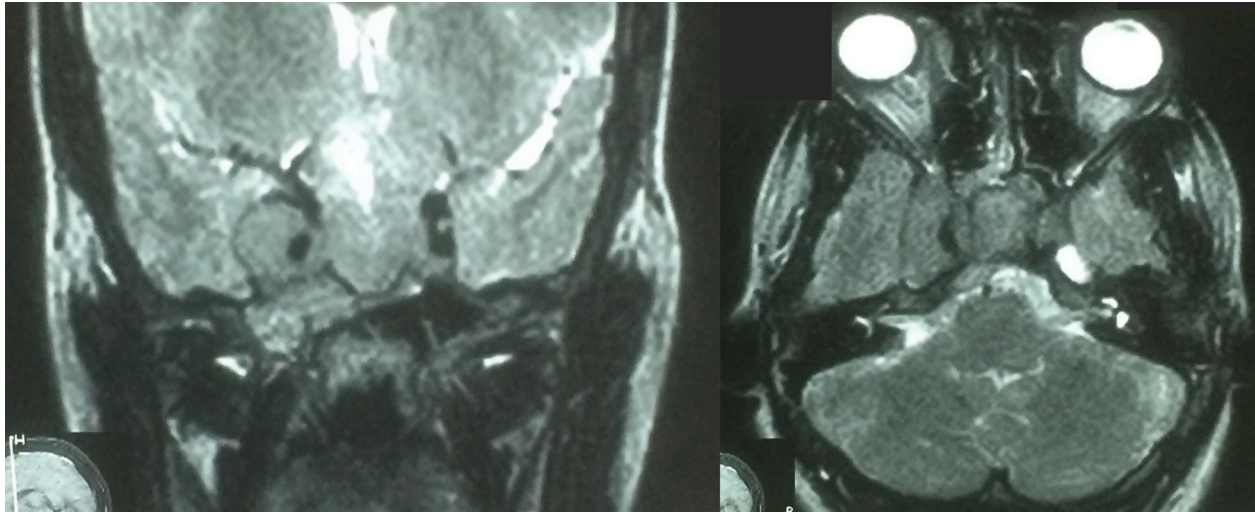


Figure 1. Brain MRI. Preoperative T2-weighted image showing total encasement of the right ICA by the pituitary adenoma. Abbreviations: ICA, Internal Carotid Artery; MRI, Magnetic Resonance Imaging.

rotation suggestive of HCHB (Video 1). When the movements were first noticed, she was on the following medications (dosage/day): levetiracetam 2,000 mg, phenytoin sodium 300 mg, cephazolin 2 g, dexamethasone 16 mg, amikacin 750 mg, and diclofenac 150 mg. Brain MRI (Figure 2) and digital subtraction angiography (DSA) (Figure 3) revealed stenosis of the supraclinoidal ICA as the cause of the infarct.

Histologic evaluation of the tumor confirmed pituitary adenoma. Her ballistic voluntary movements became more violent, leading to right upper limb abrasions. The movements occurred throughout the day and would decrease slightly when the patient was asleep. They were of moderate severity. The patient was managed by tetrabenazine 50 mg/day administered in two doses and clonazepam 2 mg in divided doses. The movements gradually reduced in severity over the next 3 months.



Video 1. HCHB. Postoperative video showing HCHB and hemiplegia on the right and left sides, respectively. Abbreviation: HCHB, Hemichorea and Hemiballism.

Discussion

Chorea is characterized by involuntary, random-appearing, irregular movements that are neither rhythmic nor repetitive. Hemichorea is a hyperkinetic disorder involving one side of the body, and ballism is defined as repetitive, constantly varying, large-amplitude involuntary movements of the proximal extremities. Hemiballism is characterized by involuntary flailing movements of the extremities on one side. If the abnormal movements are restricted to both lower limbs, it is called paraballismus; if they are generalized it is termed biballism. HCHB spans a spectrum of involuntary, continuous, nonpatterned movement involving one side of the body.

The causes of HCHB include hemorrhagic or ischemic stroke, systemic lupus erythematosus, tuberous sclerosis, hyperosmolar non-ketotic hyperglycemia, Wilson's disease, and thyrotoxicosis.² Brain lesions causing HCHB though rare, have been reported in cases of pituitary adenoma,³ putaminal cavernoma,⁴ sphenoid wing meningioma,⁵ tuberculoma,⁶ and metastatic tumor⁷ have been reported. HCHB as a surgical complication is more rare, with one reported case following surgery for craniopharyngioma.⁸ To the best of our knowledge, this is the first case reported in which HCHB developed following pituitary adenoma resection.

HCHB pathophysiology is related to direct and indirect pathways in the basal ganglia. The primary circuit of this structure is the cortico-striato-pallido-thalamico-cortical loop. Lesions in the direct pathway disinhibit the thalamus, which facilitates movements. Lesions in the indirect pathway, which includes the external globus pallidus and STN, inhibits movement. The culprit structure implicated in HCHB is the contralateral STN. Normally the STN excites the internal globus pallidus (GPi) and substantia nigra. As pallidal outflow to the thalamus is inhibitory, thalamic activity is suppressed with motor cortex activity, resulting in normal controlled movements. In HCHB, damage to the STN leads to uninhibited involuntary movements.

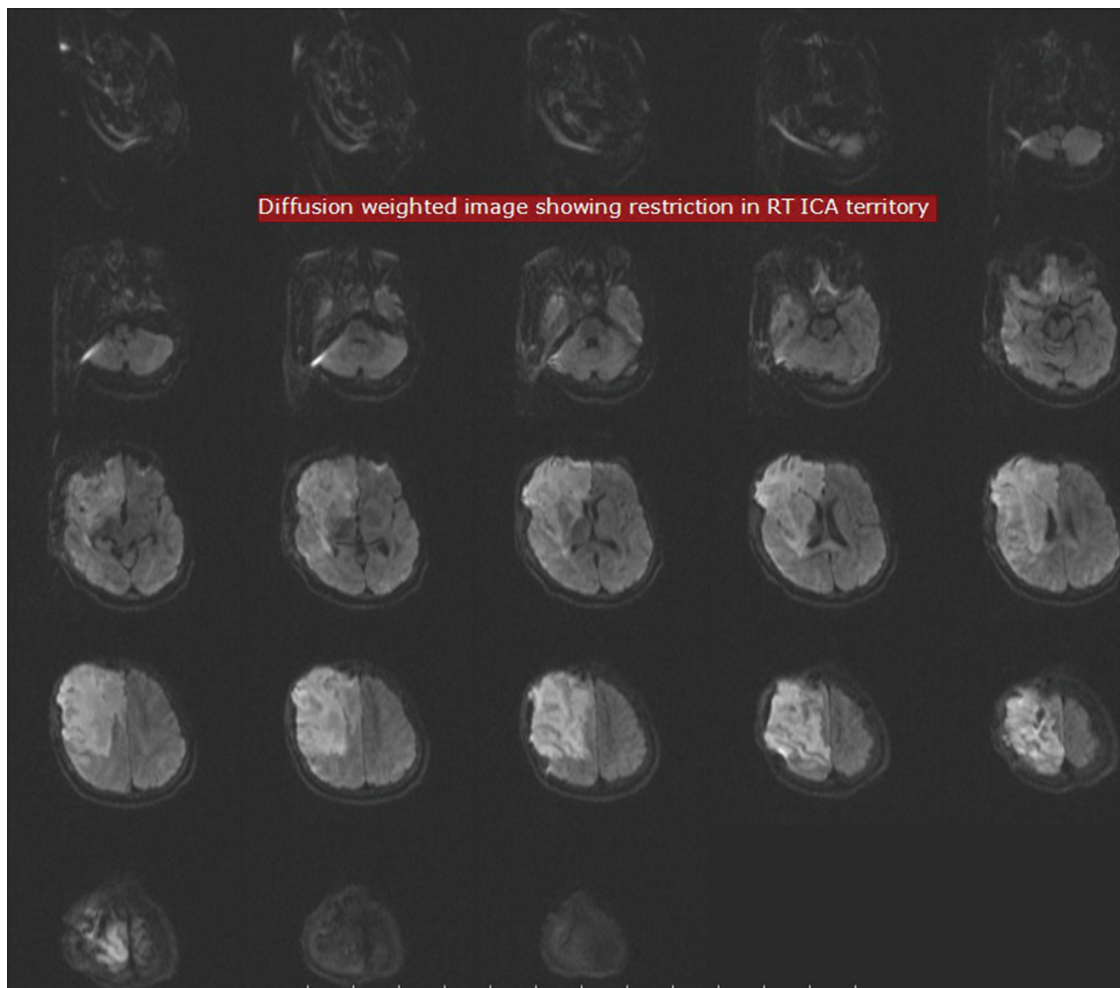


Figure 2. Diffusion MRI. Image showing restricted diffusion in the right ICA territory and sparing of the thalamus and part of the lentiform nucleus. Abbreviations: ICA, Internal Carotid Artery; MRI, Magnetic Resonance Imaging.

Ipsilateral hemiballism is rare, and the pathophysiology is unknown, although several explanations have been proposed. Only three cases of ipsilateral hemiballismus have been described. Moersch et al reported two cases following isolated ischemic lesions in the STN, whereas Borgohain et al described ipsilateral hemiballismus secondary to isolated striatal hemorrhage.^{9,10} Ipsilateral postsurgical hemiballismus was reported by Dierssen et al in a few cases of parkinsonism; however, these patients had co-existing contralateral lesions.¹¹

Biballismus with contralateral hemiplegia masking the abnormal movements is theoretically possible. However, biballismus is rare and is described in patients with multiple bilateral lesions rather than a discrete lesion in one hemisphere, as seen in the present case.¹¹

Ipsilateral HCHB could also occur when there is absence of decussation of pyramidal motor tracts. Though rare, this was reported by Hosokawa et al in a patient with ipsilateral hemiplegia caused by right internal capsule and thalamic bleeding.¹² However, our patient developed contralateral hemiplegia, ruling out this rare possibility.

Damage to the STN is classically associated with HCHB; however, lesions in other structures such as the contralateral striatum, thalamus, posterior limb of the internal capsule, and frontal and parietal cortices have also been linked to this condition. It is important to note that the ipsilateral pyramidal tract, red nucleus, pallidum, and pallidothalamic pathways must be intact to complete the circuit and manifest HCHB. In our case, diffusion MRI showed a large ICA territory infarct, indicating that areas apart from the STN are involved in HCHB pathophysiology. MRI showed that the thalamus and part of the lenticular nucleus were spared.

Based on this, a logical explanation is that there is strong disinhibition of the right thalamus due to the vascular insult, resulting in strong excitatory input to the ipsilateral cerebral cortex. The left hemiplegia suggests that the ipsilateral cerebral cortex and corticospinal tract were damaged, so excitatory output was channeled to the opposite (left) motor cortex via the corpus callosum.

It may also be noted that all ipsilateral hemiballismus cases reported to date were on the left side, with imaging showing discrete left-sided



Figure 3. DSA. Postoperative DSA showing short segment narrowing of the choroidal segment of the right ICA suggesting spasm. Abbreviations: DSA, Digital Subtraction Angiography; ICA, Internal Carotid Artery.

lesions. The present case is thus unique in that a diffuse infarct in the right hemisphere resulted in right HCHB. Thus, the possibility of a dominant left hemisphere with connections with the right basal ganglia cannot be ruled out.

The possibility that the STN exerts bilateral control over motor function may represent another explanation for ipsilateral hemiballism. This is supported by the fact that unilateral stereotaxic neurosurgery in Parkinson's disease frequently results in bilateral improvement of abnormal movements. There are no known anatomic pathways related to this bilateral control. To our knowledge, there is no evidence of crossed subthalamic projections or intersubthalamic connections.

The fact that the patient's contralateral basal ganglia and its connections were intact rules out the mechanism proposed by Dierssen et al.¹³ that highlights the importance of previous contralateral lesions in parkinsonism patients with postsurgical hemiballism.

HCHB prognosis is generally good with complete resolution in 50% of cases.¹⁴ Persistent HCHB is treated pharmacologically with neuroleptics (i.e., haloperidol, perphenazine, etc.).¹⁵ Other effective medications include tetrabenazine, reserpine, benzodiazepines, sodium valproate, gabapentin, and topiramate.^{16,17} Deep brain stimulation has been tried in refractory cases with thalamic and GPi targets.¹⁸

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