

Case Report

Hemiparkinsonism-Hemiatrophy Syndrome

Hormoz Ayromlou MD¹, Safa Najmi MD², Mohammad Ali Arami MD³

Abstract

The syndrome of hemiparkinsonism-hemiatrophy is an uncommon form of secondary Parkinsonism that presents with unilateral body Parkinsonism plus variable atrophy on the same side. Diagnosis of this syndrome needs a complete past medical history taking, as well as assessment of the familial history, clinical examination and complete paraclinical tests. The response to medical therapy has been variable in various researches. This case showed a good response to the addition of a dopamine agonist to levodopa therapy.

Keywords: dopamine agonist, hemiparkinsonism-hemiatrophy

Case report

A 34-year-old right-handed male teacher, presented with tremor for around eight years. The tremor began insidiously and progressed gradually to involve his right hand, particularly at rest. The patient had difficulty with his handwriting or with holding a cup while drinking, and general tasks that required manual dexterity. Also he felt that his balance was not quite as good as what it used to be. He has also noticed a mild twisting motion in his right hand and foot.

The patient had no history of head trauma, surgery, drug therapy, smoking, and other possible causes for movement disorders as well as relevant positive family history. He was born via natural labor without complications, but no documents exist to indicate a safe labor.

On examination, the patient had a rather regular tremor of approximately 4 to 6 cycles per second (Hz) with his right hand prominently at rest and mild dystonia in the right upper and lower limbs, particularly during working or walking. During speaking, he exhibited mild dysarthria and hypotonia. Otherwise, he had 4/5+ right hemiparesia plus mild and brief right hemiatrophy. Deep tendon reflexes were generally brisk. He had a mild bradykinesia without prominent rigidity. No autonomic problems such as orthostatic hypotension or urinary incontinence were detected. Except for mild right hemifacial atrophy, other cranial nerves were normal. His mental and cognition status, and other gener-

al and neurological exams were normal apart from a mild depression. His Unified Parkinson's Disease Rating Scale (UPDRS) scale in first visit before treatment with levodopa or other antiparkinson drugs was 43 (in the first 3 steps of the UPDRS).

Brain CT-scan showed the asymmetry between the cerebral hemispheres and ventricular asymmetry. Mild atrophy on the left hemisphere plus left lateral ventricle enlargement without other prominent lesions were also noted in the brain MRI (Figures 1 and 2).

Laboratory tests, which included FBS, hemoglobin and hematocrit, serum cholesterol, triglycerides, BUN, Cr, serum Cu, and ceruloplasmin, were all normal. Protein C and S, ANA, anti-ds-DNA, anti-phospholipids, and anticardiolipin antibodies were normal as well. Cerebrospinal fluid analysis was normal. Genetic tests for familial Parkinson (PARK1 and PARK2) were negative. No findings for motor neuron disease, neuropathies or other muscle or peripheral nervous system disorders were seen with the EMG-NC test. H/M amplitude ratio in the right extremities was also normal.

We began the patient's treatment with Rasagiline® 5 mg per 12 hours and biperidine 2 mg/8 hr because of the hand tremor, however, no improvement was noted. After three months, levodopa-carbidopa 50 mg 3 times daily was started and gradually increased to 250 mg 3 times daily over a period of three months. In contrast to previous reports, favorable results from levodopa-carbidopa alone were not seen and after five months, a dopa-agonist agent was added to the patient's treatment program after which a remarkable response was seen. Currently, the patient is on 250 mg/8 hr levodopa-carbidopa and Sifrole® 0.18 mg/8 hr plus biperidine 2 mg/8 hr.

Thus far, we have not observed any motor fluctuation or other complications during treatment. Dystonia and Parkinsonism symptoms and signs have been suppressed and cur-

Authors' affiliations: ¹Neurology Department, Imam Reza Hospital, Tabriz University of Medical Science, Tabriz, ²Neurology Department, Imam Reza Hospital, Tabriz University of Medical Science, Tabriz, ³Neurology Department, Milad General Hospital, Tehran, Iran.

Corresponding author and reprints: Mohammad Ali Arami MD, Department of Neurology Milad General Hospital, Tehran, Iran.

Fax: +98-228-222-1099; E-mail: arami_ma@yahoo.com

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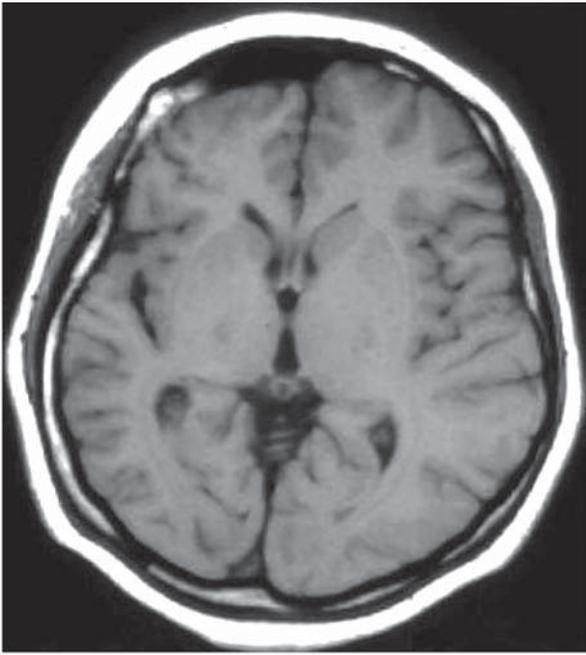


Figure 1. T1 axial plane MRI of the brain.

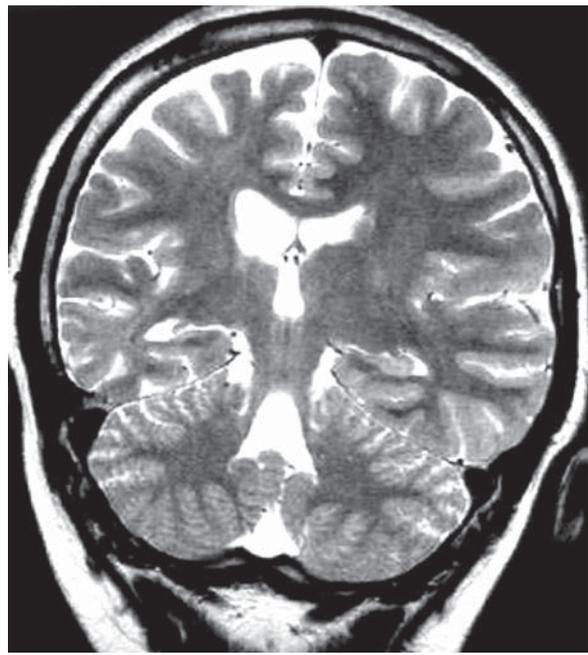


Figure 2. T2 coronal plane MRI of the brain.

rently the patient is stable and it seems that progression of his disorders has stopped. However, there are no changes in hemiatrophia. After one year of treatment his UPDRS score from the first three steps has reached 32.

Discussion

Hemiparkinsonism-hemiatropy (HPHA) syndrome is a rare form of secondary Parkinsonism that usually begins in the third or fourth decade of life; it is characterized by unilateral body atrophy and ipsilateral Parkinsonian findings with slow progression.¹ Hemiatrophy may be unnoticed.² Interestingly, one patient showed no hemiatrophy, however, an enlarged lateral ventricle was a sign of brain atrophy.³ Patients may present initially with dystonia that is often action induced and may involve only one leg or arm. There is no right or left sided prominence. Patients often have scapular winging, raised shoulder, brisk reflexes, and extensor plantars. Scoliosis is common.²

Occasionally some patients with HPHA syndrome may have problems in his fetal or perinatal periods as asphyxia or early development especially in walking.³ It has been reported that lesions of the postcentral gyrus, which occur before the age of three are associated with a relative smallness of the contralateral parts of the body.⁴ Accordingly, it has been suggested that the occurrence of Parkinsonism in HPHA patients is related to additional subcortical lesions.⁵ Similarly, Giladi et al. have presented neuroradiological

evidence of contralateral brain hemiatrophy in 64% of their patients. Since an association between perinatal asphyxia, brain hemiatrophy, and delayed-onset hemidystonia has been shown, therefore HPHA could represent an example of a movement disorder with delayed onset as a consequence of neonatal brain injury.³

CT-scan and MRI in one study showed contralateral brain asymmetry in 64% of investigated patients,³ but Bushman et al. found no evidence of cerebral hemiatrophy.⁶ Giladi et al. have reported 11 cases of HPHA in which 6 patients presented with body and contralateral cerebral hemispheric hemiatrophy as seen on brain imaging, 4 with solely body hemiatrophy and one with only brain hemiatrophy. MRI findings were asymmetric lateral ventricles, volume loss, thalamic, or arachnoid cysts. Unilateral changes included calvarial thickening, expansion of the ethmoid, frontal sinuses, or mastoid cavities, and elevation of the petrous ridge and greater wing of the sphenoid bone.² Studies that utilized 18F-fluorodeoxyglucose and positron emission tomography (PET) have shown focal hypometabolism in the basal ganglia and the medial frontal cortex of the contralateral side.⁷

In contrast to presynaptic involvement in parkinsonism's disease (PD), PET scanning have provided evidence of presynaptic and postsynaptic nigrostriatal dopaminergic dysfunction in patients with HPHA syndrome.^{8,9} One patient with HPHA syndrome was found with a mutation in the Parkin gene on chromosome 6.¹ Negative signs and symptoms that are rare or not seen at all include axial signs such

as swallowing or speech difficulty or symmetric freezing.²

Responses to the drug therapy with levodopa vary among studies. In case reports from Giladi et al., three patients showed good response to levodopa (33%), four had moderate response (44%) and two patients (22%) had a poor response.³ Bushman et al. have presented the HPHA syndrome as a dopa-responsive condition.⁶ Wijmann et al. reported good response to levodopa in 18 (60%), moderate in 6 (20%), and poor in 6 cases (20%).² Jenkins et al. reported a case of HPHA syndrome that showed dramatic improvement with subthalamic nucleus stimulator (STN) implantation.¹⁰ However, the effects of dopamine agonists in treatment of HPHA syndrome are not clear due to the low numbers of cases and lack of published results with these agents. We have used these agents based upon their therapeutic effects in idiopathic Parkinson's disease. As a result of the good response of our patient, therefore we recommend this combination therapy in levodopa-resistant cases of HPHA syndrome.

Up to the knowledge of authors, only few cases of this disease have been reported worldwide, and its real pathophysiology is not clearly known.

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Gardenehye Ghoochak, around 8 Km from Tehran, winter 2011. (Photo by M. H. Azizi MD)