

by Kozłowski et al² and of our work is that heterotopic neurons appear resistant to the degenerative process that affects motor neurons in ALS, while other spinocerebellar neurons are frequently involved in the disease.⁸⁻¹⁰ Averbach and Crocker⁸ found that more than one-third of neurons in Clarke's nucleus were lost in patients dying with ALS, a finding confirmed by Williams et al¹⁰ in a study using a monoclonal marker for neurons together with another monoclonal antibody that reacted more specifically for a surface marker on spinocerebellar neurons in Clarke's dorsal nucleus and in the neurons of the cervical spinocerebellar and ventral spinocerebellar tracts. The lack of involvement of neurons located in the white matter of the spinal cord in ALS, SMA, and poliomyelitis suggests that these neurons are not part of the motor or spinocerebellar systems and that they may be sensory neurons, despite their frequent location in the ventral part of the white matter of the cord.

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Hemiballismus and secondary mania following a right thalamic infarction

Article abstract—An 81-year-old woman had the sudden onset of left-sided ballismus and an accompanying behavioral change characterized by elation, distractibility, inflated self-esteem, and insomnia, suggesting secondary mania. An MRI revealed a small ischemic infarction of the right thalamus.

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There are only occasional reports of mental changes associated with hemiballismus.¹ We describe a patient who presented with acute hemiballismus and a disinhibition syndrome mimicking mania after a small right thalamic infarction.

Case report. An 81-year-old, right-handed woman suddenly developed involuntary movements of the left limbs. During the next 3 days, her relatives noted a marked change in her behavior. She became increasingly euphoric and talkative, and had grandiose delusions. She was admitted to hospital on the fourth day. She had diabetes and well-controlled hypertension but no familial or personal history of affective disorders.

On hospital admission, she was globally oriented, but appeared distractible and had pressured speech and

flight of ideas. She believed that her health was better than ever and joked inappropriately. She also reported a decreased need for sleep. Her mood changes, which were assessed with the Structured Clinical Interview from DSM-III-R,² met criteria for an organic affective disorder, manic type. On a mania rating scale,³ her mania score was 15 points (definite mania). She scored 26 out of 30 on the Mini-Mental State Examination.⁴ On neurologic examination, she showed choreic and ballistic movements of the left limbs. There were almost-constant, small-amplitude, proximal and distal choreic movements, with superimposed brisk, large-amplitude proximal movements, primarily related to action. Sensation, strength, and tendon reflexes were normal. Plantar responses were flexor bilaterally. Ocular motility and visual fields were normal, and there was no evidence of visual neglect on a

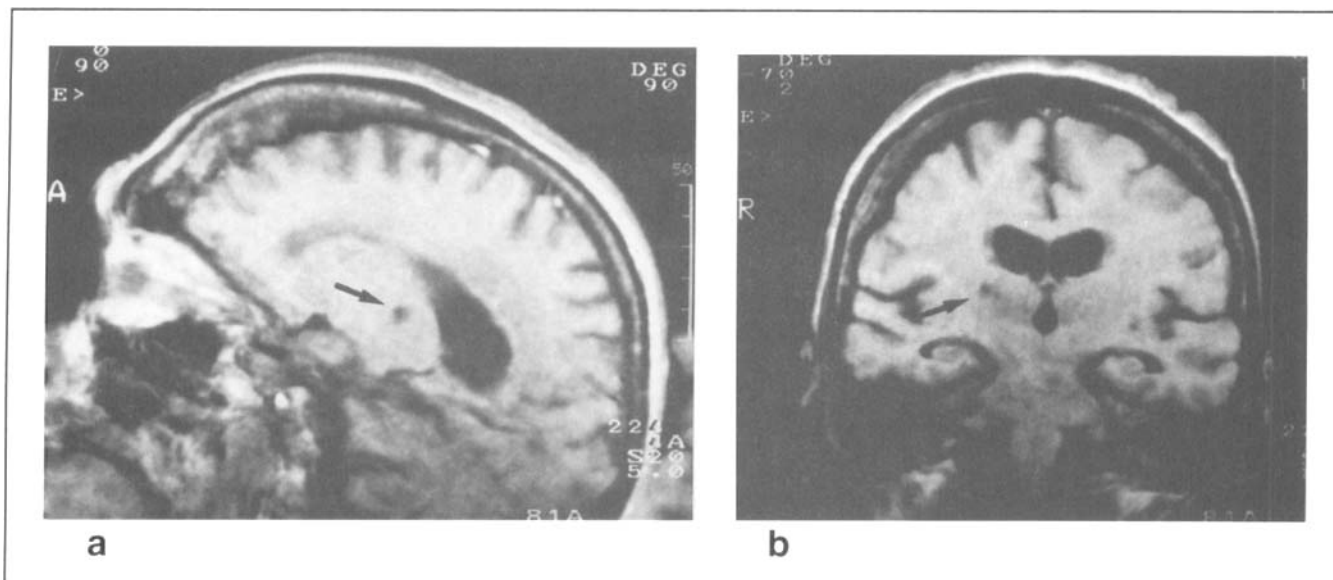


Figure. T_1 -weighted MRI ($TR = 2,000$; $TE = 40$) showing a hypointense thalamic lesion (arrows) at the level of the right ventral lateral and ventromedial nuclei in (panel a) the sagittal (A = anterior) and (panel b) the coronal planes (R = right).

line crossing-out task. An MRI with 5-mm sections revealed a small area of hypointense T_1 signal in the right thalamus (figure). The lesion involved the ventral lateral and ventromedial nuclei but spared the cerebral peduncle and midbrain. The patient was started on haloperidol (15 mg/d), and the motor and behavioral disturbances improved by the third day. Affective disturbances subsided 1 month after the onset, and haloperidol was then gradually discontinued. At 3 months follow-up, the patient remained euthymic, but she still showed mild choreic movements of the left limbs.

Discussion. Although most cases of hemiballismus result from small vascular lesions involving the contralateral subthalamic nucleus or its projection target areas, lesions of the contralateral thalamus may produce similar motor symptoms.⁵ There are occasional reports of mental changes in patients with hemiballismus, particularly with lesions extending beyond the subthalamic nucleus.^{1,5} In these cases, hemiballismus and behavioral changes generally coexisted with autonomic and sensory symptoms and altered strength, muscle tone, stretch reflexes, or other clinical features suggesting large lesions.^{1,5} Our case is unique in that hemiballismus and secondary mania occurred without concomitant neurologic manifestations, and the responsible lesion was small and restricted to the right thalamus.

Secondary mania after thalamic damage was reported occasionally in association with ischemic and hemorrhagic lesions, neoplasms, sarcoidosis, stereotactic thalamotomy, Kleine-Levin syndrome, or as a postoperative complication in coronary artery bypass surgery.⁶⁻⁸ The secondary mania after a right thalamic lesion is in accord with previous reports implicating dysfunction of limbic and limbic-connected areas (ie, orbitofrontal or basotemporal cortices, caudate nucleus, and thalamus) of the right hemisphere as a cause of organic

mania.^{6,7} The right thalamic infarction may have selectively disrupted the convergence of limbic and subthalamic pathways to various thalamic nuclei as well as the thalamocortical connectivity.

The coexistence of mania and movement disorders was reported in a number of basal ganglia disorders, specifically Huntington's disease, Sydenham's chorea, postencephalitic parkinsonism, Wilson's disease, and Tourette's syndrome.^{8,9} While episodes of mania are relatively frequent in patients with hyperkinetic movement disorders such as early Huntington's disease, they are uncommon in patients with hypokinetic movement disorders such as Parkinson's disease. In Parkinson's disease, manic states and ballistic movements similar to those seen in our patient were most often reported in patients with levodopa-induced "on-off" motor fluctuations. In these patients, manic swings and dyskinetic movements mainly occurred during "on" periods and were attributed to striatal and limbic dopaminergic overactivity.¹⁰ An analogous dopaminergic overactivity is postulated in Tourette's syndrome, a condition in which a disinhibition syndrome is frequently seen.⁹ Therefore, it is possible that a similar biochemical mechanism could be implicated in the pathogenesis of mania and hemiballismus in our patient. In support, haloperidol, a postsynaptic D_2 -receptor antagonist, resulted in suppression of mania and hyperkinetic movements.

Our patient's improvement with haloperidol is consistent with the literature⁷ in which reported cases with secondary mania had good response to treatment with the usual antimanic drugs—lithium or neuroleptics. Perhaps prompt treatment of hemiballismus patients with neuroleptic therapy masks accompanying behavioral changes, such as mania.

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Familial band heterotopias simulating tuberous sclerosis

Article abstract—We report the clinical and neuroimaging findings of a mother and daughter with seizure disorders and band heterotopias seen on magnetic resonance imaging studies. These clinicoradiologic findings simulate those for a diagnosis of tuberous sclerosis complex. Clinicians should be aware of this migrational anomaly and its neuroimaging characteristics, as well as the potential for this specific migrational anomaly to be genetically transmitted.

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MRI allows the appreciation of more detailed in vivo neuroanatomy. A number of neuronal migrational abnormalities have become more easily recognized as compared with conventional CT scanning.¹⁻³ A recently recognized migrational anomaly, band heterotopias, bears a resemblance to the subependymal nodules (SEN), a migrational anomaly seen in patients with tuberous sclerosis complex (TSC).^{4,6} We report a 16-year-old patient and her mother, both with band heterotopias, and describe how this syndrome is distinguishable from TSC and SEN.

Case report. A 16-year-old woman of Native American and German extraction with a mild seizure disorder was referred to the neurogenetics clinic at the University of Connecticut for evaluation of possible tuberous sclerosis. Referral was prompted by the recognition of periventricular heterotopias on the head CT.

She was born after an uneventful pregnancy at full term, attaining appropriate developmental milestones. She had a first and second generalized tonic-clonic seizure associated with fever between ages 3 and 5 years. Neurologic and general examinations as well as an EEG at that time were normal. A head CT reportedly showed enlarged ventricles. Intermittent phenobarbital was administered but not maintained. At age 10, she experienced her first nonfebrile generalized seizure. A repeat EEG demonstrated bicentral paroxysmal activity with

right-sided predominance and generalized paroxysmal discharges during sleep. A head CT at this time demonstrated enlarged ventricles and periventricular irregularities subsequently defined as heterotopias. Carbamazepine was prescribed after a second nonfebrile, complex partial seizure. The frequency of seizures was limited to isolated yearly complex partial events during the first 3 years of therapy and remitted over the past 2 years. She remains an excellent student. Her past history is otherwise unremarkable. The parents denied consanguinity.

The patient's 35-year-old mother was diagnosed with complex partial seizures at age 21 after an evaluation for recurrent episodes of right arm numbness, aphasia, and altered consciousness. Her head CT was normal, and she was eventually treated with carbamazepine and phenobarbital. Seizure frequency is about twice per year. The maternal grandmother, now deceased, also had epilepsy requiring therapy with phenytoin. Details of her clinical examination are lacking, but she did not have renal problems or obvious facial angiofibroma. The remainder of the family history was noncontributory.

On examination under Wood's lamp, the patient had a difficult-to-recognize single ill-defined hypopigmented macule and a small café au lait spot on the right flank. There were no facial angiofibromas, poliosis, confetti lesions, gingival or ungual fibromas, connective tissue nevi, collagen plaques, or dental pits. Dilated funduscopy was normal. The remainder of her general and neurologic examinations were normal. Subsequent brain MRI

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