Hemianopsia in Dementia With Lewy Bodies

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Objective: To describe the pathologic changes that caused a left homonymous hemianopsia in a patient with dementia with Lewy bodies.

Design: Report of a case and postmortem studies.

Main Outcome and Results: A 66-year-old woman experienced parkinsonism and left homonymous hemianopsia early in the course of a rapidly progressive dementia that culminated in death only 21 months after the onset of her symptoms. Postmortem examination revealed pathologic features consistent with the diagnosis of dementia with Lewy bodies. The only apparent explanation for her visual field deficit was a disproportionately large number of neurofibrillary tangles in the right striate, peristriate, and inferotemporal cortices.

Conclusion: A clinically obvious homonymous hemianopsia can result from the occipital and inferotemporal cortical degeneration in dementia with Lewy bodies.

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Dementia with Lewy bodies has an uncertain nosology and a varied clinical terminology, which includes diffuse Lewy body disease, Lewy body dementia, dementia with cortical Lewy bodies, senile dementia of Lewy body type, and Lewy body variant of Alzheimer disease. The term dementia with Lewy bodies (DLB) was recently proposed by the Consortium on Dementia with Lewy Bodies and is used in this article. Most patients exhibit a combination of dementia, parkinsonism, and psychiatric symptoms, but striking deviations from this symptom complex are common, leading to a variety of incorrect clinical diagnoses. Herein we report a unique patient with DLB who initially complained of heaviness in the right upper extremity and subsequently developed a dense left homonymous hemianopsia during the course of a rapidly progressive dementia.

REPORT OF A CASE

A previously healthy 66-year-old right-handed woman was hospitalized with a 5-day history of heaviness in her right upper extremity and a flulike illness, associated with malaise, nausea, vomiting, and diarrhea. She attributed her right upper extremity symptoms to an unusual amount of writing done while completing her income tax returns. Her internist suspected a stroke and performed a computed tomographic scan of the head, which revealed a 6-mm lucency in the deep left frontal white matter. Magnetic resonance imaging revealed a 6-mm subcortical focus of increased T2 signal in the same location. A neurologist (R.J.E.) found that she was a cooperative historian who scored 26 of 30 on the Folstein Mini-Mental State Examination. She remembered 0 of 3 words at 1 minute and did a very poor job of copying intersecting pentagons. She had difficulty spelling the word “world” backward, but she corrected herself and eventually accomplished this task. She was able to pantomime simple motor tasks. There was no aphasia. Cranial nerves were normal, including voluntary eye movements and optokinetic nystagmus. She exhibited mild facial masking, hypophonia, micrographia, and cogwheel rigidity in both upper extremities, greatest in the right. She had no tremor or weakness, and muscle stretch reflexes, plantar responses, and sensory examination were normal. Arm swing while walking was diminished bilaterally, greatest on the right, and her steps were slightly short and cautious but not shuffling. Postural stability was nor-
mal. An outpatient neurologic examination 4 weeks later revealed the same motor signs but an improved Mini-Mental State Examination score of 29. She lost 1 point because of her persistent inability to draw intersecting pentagons (Figure 1).

Eight months later, she returned to the neurologist with the complaint of reduced vision in her left eye. This developed insidiously, and she was unsure of its duration. Examination revealed an obvious left homonymous hemianopsia to finger counting and gross hand waving. This visual deficit was present in both eyes and was particularly dense in the inferior quadrant. Her neurologic examination and Mini-Mental State Examination score were otherwise unchanged. An ophthalmologist found no significant ocular pathologic conditions (retina, optic disc, and anterior chamber). Humphrey automated perimetry was somewhat limited by her poor attention, but the results were consistent with a left homonymous hemianopsia (Figure 2). Another magnetic resonance imaging scan performed with and without contrast was unchanged since the initial study.

A gradual severe deterioration in cognitive and motor function occurred during the next 12 months. She had periods of severe confusion, agitation, depression, and visual hallucinations of animals and unfamiliar people in her home. In desperation, she was taken to several other physicians and was treated unsuccessfully with selegiline hydrochloride, a combination product of carbidopa and levodopa, vitamin E, thiroidazine, alprazolam, fluoxetine hydrochloride, thiothixene, benztropine mesylate, trazodone hydrochloride, lorazepam, and electroconvulsive therapy. The trial of electroconvulsive therapy was aborted after 1 treatment because of severe delirium. Administration of thioidazine (50 mg/d) and thiothixene (5 mg/d) each produced a disabling increase in her parkinsonism. Selegiline hydrochloride (5 mg) administered twice daily and a combination prod-
COMMENT

Our patient exhibited progressive cognitive decline, parkinsonism, visual hallucinations, and fluctuating agitation, confusion, and depression. She therefore fulfilled all the consensus criteria for the clinical diagnosis of probable DLB.7,8 Dementia with Lewy bodies was our initial diagnosis, but the rapid progression with myoclonus and the left homonymous hemianopsia were cause for continued diagnostic uncertainty. Initially, the asymmetric motor signs and visual field deficit led to an unproductive search for cerebral vascular disease. The erroneous clinical diagnoses that were considered over the course of this patient’s 21-month illness included stroke, Parkinson disease, vascular dementia,Binswanger disease, major depression, Alzheimer disease, and Creutzfeldt-Jakob disease. Such diagnostic uncertainty is common in individuals with DLB because of its varied clinical picture and course.1-7

The neuropathologic findings in our patient fulfilled the diagnostic criteria for DLB, proposed by the Consortium on Dementia with Lewy Bodies.7,8 The pathology in our patient was similar to that in other patients with DLB,3-5,7,8 except our patient exhibited a striking predominance of neurofibrillary tangles in the right inferotemporal and occipital cortices. Alzheimer pathologic manifestations occur in most cases of DLB, but neocortical tangles are seen in a minority of patients with DLB.7 The high density of tangles in the right occipital and inferofrontal cortex was the only apparent explanation for our patient’s early constructional apraxia and symmetric left homonymous hemianopsia.

Did our patient have both Alzheimer disease and DLB? This question is unanswerable because the distinguishing clinical and neuropathologic criteria are still being defined.7 The Consortium on Dementia with Lewy Bodies2 acknowledged that Alzheimer pathologic features are common occurrences in DLB, and our patient

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illustrates the difficulty in separating Alzheimer disease from DLB. Her relatively young age makes age-associated Alzheimer features an unlikely explanation for the Alzheimer pathologic changes in her brain.

An inferotemporal-occipital predominance of tangles is not characteristic of Alzheimer disease or DLB, but such pathologic changes are found in patients with Alzheimer disease with unusually severe visual impairment.10-15 The visual deficits in these patients with Alzheimer disease include Balint syndrome, constricted peripheral visual fields, visual agnosia, quadrantanopsia, and hemianopsia.10-15 Quadrantanopsia and hemianopsia are rare in individuals with Alzheimer disease12-14 and have not been reported previously in individuals with DLB. Furthermore, we know of no other case in which quantitative neuropathologic examination was performed to explain the hemianopsia. Previous patients with Alzheimer disease with quadrantanopsia and hemianopsia had coexistent simultanagnosia, but the hemianopsia preceded the simultanagnosia in our patient, consistent with her markedly asymmetric inferotemporal-occipital pathology.

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