SHORT REPORT

Slowly progressive apraxia in Alzheimer’s disease

R C Green, F C Goldstein, S S Mirra, N P Alazraki, J L Baxt, R A E Bakay

Abstract

Slowly progressive apraxia due to Alzheimer’s disease was encountered in a 66 year old, right handed man whose initial impairments included coordinated movements of the left hand and some features of the alien hand syndrome. Over four years, the patient developed progressively worsening deficits of memory and language. A biopsy of his right temporal lobe showed numerous plaques and neurofibrillary tangles. Pronounced right parietal lobe hypoperfusion on serial SPECT suggests involvement of this region in contralateral praxis.

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Keywords: Alzheimer’s disease; subtypes; apraxia; movements

Apraxia is a syndrome of impaired execution of learned skilled movements that cannot be explained by weakness, incoordination, sensory loss, lack of comprehension, or inattention. Apraxia is common in Alzheimer’s disease, usually appearing after impairments of memory and language are established. A few cases of slowly progressive apraxia without deficits in memory and language have been described but the underlying pathology was not established. Our patient showed progressive apraxic symptoms early in the course of his illness.

Patient history

A 66 year old, right handed, college educated chemical engineer experienced the insidious onset of finger and hand incoordination in association with complex movements of the left hand at the age of 60. Initially, he had minor problems driving his car and shuffling playing cards, but by the end of 1988 he had stopped working and could not easily tie his shoelaces, button his shirt, or eat with his left hand. He and his family denied any language or memory difficulties. When initially examined in June 1990 the patient was aware of his deficits, stating that his left hand “seems to have a life of its own.” His wife confirmed extraneous behaviours reminiscent of alien hand syndromes, such as placing a napkin in his lap with his right hand, only to have his left hand reach out and remove it. After six months, he had problems performing mental arithmetic, writing script or numbers with his right hand, and he experienced spatial confusion when driving or in a familiar environment without visual cues. He and his wife agreed that he still did not have problems with gait, word finding, comprehension, or memory. There was no personal or family history of neuropsychiatric problems or substance misuse, except for a maternal aunt clinically diagnosed as having Alzheimer’s disease. Detailed neurological examination was normal, including motor tone and primary sensory and motor function in the limbs, except for moderately severe agaphaesthesia and atetognosia in the left hand and difficulties with left hand praxis.

Neuropsychological evaluations were conducted in July 1990, January 1991, and July 1992 (table). Apraxia was evaluated by having the patient carry out gestures and imagined use of objects. When seen in July 1990, imitation was performed poorly by each hand and the execution of transitive and intransitive movements to command was worse with the left hand. Responses were slow, and body part as the object and spatial displacements were common. Performance did not appreciably improve when the patient was tested with actual objects. At his six month follow up, there was a noticeable deterioration of transitive and intransitive movements with the right hand. Imitation was still poor bilaterally, and had become even worse with the left hand. By July 1992 he could only perform simple right handed movement to command and imitation, and he could not execute any movements with his left hand.

In July 1990 he could not determine how to hold a pencil with his left hand and he could not write a sentence correctly to dictation with his right hand, although he could copy. By January 1991, his writing with his right hand, both to dictation and from copy deteriorated and by July 1992 he was unable to write his name. His drawings were tremulous and micrographic, and he was impaired in copying simple geometric designs on the Benton visual retention test. By contrast with his dyspraxic and dysgraphic symptoms, his language abilities were relatively preserved in July 1990 and January 1991. By July 1992, however, he displayed severe dysnomia, with many perseverual misidentifications and semantic paraphasias. Generation of words
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Neuropsychological test results

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<tr>
<td>Intellectual function</td>
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</tr>
<tr>
<td>Verbal IQ</td>
<td>109</td>
<td>110</td>
<td>69</td>
</tr>
<tr>
<td>Performance IQ</td>
<td>63</td>
<td>60</td>
<td>60</td>
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<tr>
<td>Full Scale IQ</td>
<td>88</td>
<td>87</td>
<td>63</td>
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<tr>
<th>Verbal age scaled scores: (mean = 10, SD = 3)</th>
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<tbody>
<tr>
<td>Information</td>
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<tr>
<td>Digit span</td>
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<tr>
<td>Vocabulary</td>
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<td>Arithmetic</td>
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<td>Comprehension</td>
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<td>Similarities</td>
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<th>Performance age scaled scores:</th>
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<tr>
<td>Picture completion</td>
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<tr>
<td>Picture arrangement</td>
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<tr>
<td>Block design</td>
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<td>Digit symbol</td>
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Language:

Apraxia (No correct)
- Limb intransitive (12 total):
  - RH: commands
  - LH: commands
  - Imitation
  - Limb transitive (7 total):
  - RH: commands
  - Imitation
  - LH: commands
  - Imitation

Writing (No correct/6 words)
- Dictation (RH, LH)
- Copy (RH, LH)
- Naming (No correct/60 pictures)
- Verbal fluency (No correct)
- Three letters
- Three categories
- Tactile naming (14 objects)
  - RH (No correct)
  - LH (No correct)

Memory:

Picture recognition (No correct/100)
- Story recall (No units/20)
- Story recognition (No units/20)

CND = could not do; NG = not given; LH = left hand; RH = right hand.

Propylene amineoxime with the patient supine in a low stimulation environment in August 1990 and February 1991. These studies showed hypoperfusion in the right frontal, parietal, temporal, and occipital regions (fig 1).

In February 1991, biopsies of the middle and inferior temporal cortices on the right side were performed. Neuropathological examination with Bielschowsky silver staining of the paraffin embedded material showed numerous senile plaques (up to 50 per low power field) (fig 2). The plaques were predominantly neuritic in nature, some containing amyloid cores; a few diffuse plaques were also seen. The density of neuritic plaques met conventional neuropathological criteria for diagnosis of Alzheimer’s disease. Numerous neurofibrillary tangles were also noted, particularly in the deeper layers of cortex, along with neuropl threads.

Discussion

Apraxia is common in dementia, occurring in Huntington’s disease, corticobasal ganglionic degeneration, and occasionally in Parkinson’s disease. Apraxia is also common in Alzheimer’s disease, but is rarely described as a presenting symptom. Three cases of slowly progressive apraxia without tissue diagnoses have been reported, but two of these seemed to represent visual agnosias in which the patients made errors imitating gestures but performed correctly to command. The third case, a patient with an insidious loss of abilities to perform limb and axial movements to both imitation and command, more closely resembles our patient, but there was no follow up or tissue diagnosis.

Patients with Alzheimer’s disease established by biopsy have presented with progressive left sided clumsiness, asterognosia,agraphaesthesia, and choreoathetoid movements as well as with alien left hand and myclonus. Several other recent clinical case reports of slowly progressive apraxia have been associated with unilateral or bilateral cortical atrophy and diminished SPECT activity in parietal regions but none of these reports included a tissue diagnosis. Published reports of SPECT findings in patients with Alzheimer’s disease indicate that parietal lobe hypoperfusion is often seen, but it usually presents bilaterally. Unilateral apraxia has often been described in patients with focal lesions, particularly strokes, but is also unusual in Alzheimer’s disease.

Apraxia of the non-dominant limb may be a result of interhemispheric disconnection associated with lesions of the corpus callosum fibres, and is often, but not exclusively, associated with left hemispheric lesions and with aphasia. Although it is less frequent, apraxia can also occur after right hemispheric lesions, raising the possibility that the visuokinaesthetic engrams containing spatial and temporal representations of learned movements may occasionally be represented either bilaterally or in the non-dominant
hemisphere, and not only in the left hemisphere.20-24

There is no way of knowing the full extent of neuropathology in a living patient with neurodegenerative disease. It is of interest, however, that this patient presented with prominent symptoms of the left hand and that SPECT perfusion images and quantitative analysis showed mild frontal and pronounced parietal hypoperfusion in the right hemisphere, supporting the notion that these regions may be salient in contralateral praxis. Indeed, the praxis system in the right hemisphere has been implicated in the mediation of “concrete” or context dependent performance of transitive movements and in learning novel movement sequences.25 The alien hand features seen in this patient, along with dysgraphia and impaired tactile naming with the left hand, are difficult to explain on the basis of frontal or parietal lesions alone and suggest some degree of interhemispheric disconnection early in the disease process.

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18 Halvand KY, Flaherty D. The different types of limb apraxia errors made by patients with left vs right hemisphere damage. Brain Cogn 1984;3:370–84.

NEUROLOGICAL STAMP

Chaulmoogra (Hydnocarpus wightiana)

The seeds of the fruit of the chaulmoogra tree contain strongly antibacterial chemicals, two of which, hydno-carpic and chaulmoergic acids, destroy the bacterium Mycobacterium leprae. Ancient Hindu and Chinese documents described an oil that was effective against leprosy, and it is likely that this came from the chaulmoogra tree. Only about the middle of the last century was the oil taken seriously by western physicians. It was investigated, tested, and soon imported from China but the supply was severely limited. In some places today, the ingredients of chaulmoogra oil, modified by chemists, are still used to cure early cases of leprosy, but advanced cases do not yield to the treatment.

The chaulmoogra tree is shown on a stamp issued by Fiji in 1970 (Stanley Gibbons 420, Scott 289).

L F HAAS
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