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SLOWLY PROGRESSIVE APHASIA FOLLOWED BY ALZHEIMER'S DEMENTIA

A CASE REPORT

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SUMMARY — Slowly progressive aphasia has been found in 8 published cases, 2 of whom progressed over a period of years to generalized dementia. Positron emission tomography demonstrated decreased glucose metabolism in the left perisylvian region in 2 cases. We describe a patient who had slowly progressive aphasia and developed generalized Alzheimer's dementia 7 years after presentation. There was no clinical or laboratory evidence of concomitant disease. Computerized tomography showed generalized atrophy more marked on the left perisylvian region late in the disease, when EEG showed generalized slowing more marked on the same area. Slowly progressive aphasia of old age should be considered a separate entity until further studies elucidate its relation to Alzheimer's dementia.

Afasia lentamente progressiva seguida de doença de Alzheimer: relato de um caso.

RESUMO — A literatura registra 8 casos publicados de afasia lentamente progressiva, dois dos quais progrediram em período de anos para demência generalizada. Tomografia por emissão de pósitrons demonstrou metabolismo diminuído de glicose na região peri-silviana esquerda em dois casos. Descrevemos o caso de um paciente que teve afasia lentamente progressiva e desenvolveu demência generalizada de tipo Alzheimer, 7 anos após a apresentação inicial. Não foi encontrada evidência clínica ou laboratorial de doença concomitante. Tomografia computadorizada mostrou atrofia generalizada mais marcada na região peri-silviana esquerda tardiamente no processo, quando EEG mostrou lentificação generalizada, mais claramente na mesma área. Afasia lentamente progressiva da idade avançada deve ser considerada entidade em separado até que estudos elucidem sua relação com demência de Alzheimer.

Focal hemispherical syndromes may overlap with generalized dementias. Ron et al.[7] found 31% of patients previously diagnosed as demented to have another diagnosis. Focal neuro-psychological syndromes may produce well-defined deficits of cognitive and intellectual functions which may be confused with dementia [4,5]. Patients with aphasia are specially likely to be erroneously considered as demented in view of the difficulties in communication which may make a complete diagnosis of other aspects of mental function almost impossible [4,6,9]. Language deterioration in Alzheimer's disease starts with difficulty in the correct use of words and circumlocutions. Later there may be frank anomia and paraphasia, echolalia, dysarthria or mutism [3]. Although similar to aphasia due to a focal progressive lesion, the diagnosis of Alzheimer's disease is strongly suggested by the concomitant disintegration of other hemispherical functions [5].

We report a patient who had a slowly progressive aphasia which 7 years after onset, when very severe, became accompanied by generalized dementia. We have found other similar cases published by the same group in the American literature [1,6].

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CASE REPORT

IDU, a 70-year-old man at the time of writing (March 1987) is a retired right-handed white caucasian lawyer who throughout his life was an avid reader of his professional literature, of literature in general and of daily newspapers. He was successful in his career and kept well informed about the general world situation. He had a regular life, did not smoke or drink alcohol. One brother is a 74-year-old retired army general, developing symptoms of classical Alzheimer's dementia at the time of writing, after 2 years of a depressive behavioural change. Other 6 brothers and sisters aged between 67 and 79 years have not shown signs of dementia or of progressive aphasia according to family information. The patient was first seen by one of us (PRMB) in 1982 with a 2-year history of difficulty in the comprehension of written texts. His wife reported some apathy but his behaviour, intellect and general health were otherwise quite normal and he was able to locate himself within his house, to keep a high degree of hygiene and elegance, to do all the house shopping and look after his investments and businesses. On examination there was no ataxia, there was little mobility of the face, and smooth pursuit eye movements were abnormaly broken. Visual field, pain and position sense were difficult to evaluate. Comprehension of isolated sentences, naming of objects and the ability to carry out simple calculations were preserved, but during conversation and on complex commands decreased motor output and some difficulty in understanding verbal information were noted. Comprehension of written texts was impaired to the extent that he had difficulty reading aloud or explaining the meaning of newspaper headlines. The deficit was more pronounced with magazines or newspapers texts. He was able to copy written words and numbers. Performance on dictation and spontaneous writing were in accordance with his intellectual state. Routine blood examinations, computerized axial tomography (CAT) and EEG were normal. The patient was referred to speech therapy.

When seen by the speech therapist (MJM de OC) in 1984 the patient was noted to keep quiet during conversation, answering with short phrases and low voice when questioned. He and his wife reported a clear progress in the reading deficit specifically with regard to understanding newspaper texts which he was used to read since youth. Examination showed difficulties in writing with hesitation especially in spontaneous writing and in a lesser degree during dictation, although copying was intact. The patient was alert, lucid and maintained his personal hygiene, elegance, house shopping and businesses with his usual methodic characteristic. He was able to attend speech therapy rather far from his residence, involving 2 bus rides and a certain amount of walking. When seen in 1985 there was clear difficulty in naming objects, difficulty with writing during dictation with paragraphia, difficulty in simple calculations and complete inability to read. The patient kept his usual pace of life except for some difficulty with his businesses and bank accounts, which could be ascribed to his speech problems.

In 1986 he was seen for the second time by a neurologist (PRMB) and admitted for further studies. At this stage he rarely uttered isolated syllables or obscure rumbles but he was able to repeat simple words quite easily. Comprehension of verbal commands was moderately impaired. He did not read, did not write spontaneously, did not perform simple calculations, did not understand written material, but he was able to copy with some difficulty drawings of objects, some simple words and numbers. General physical examination was normal, specifically with regard to vascular, inflammatory or neoplastic disorders. Other cortical and behavioural functions were preserved except for a certain degree of apathy. Visual field and sensory testing were not reliable. The remaining of the neurological examination was normal. Full blood count, ESR, blood sugar, syphilis serology, creatinine, chest X-ray and ECG were normal. The cerebral spinal fluid was normal except for a protein of 54 mg/%. EEG showed slowing of the dominant rhythm to between 5 and 6 cycles per second, more severely on the left. There were diffuse slow waves between 4 and 5 cycles per second as waves between 1 and 2 cycles per second of 10 to 100 microvolts localized on the left fronto-temporo-parietal region, noted to occur in short bursts. CAT showed evidence of generalized cerebral atrophy perhaps more marked posteriorly on the left hemisphere (Fig. 1). The patient was discharged without a definite diagnosis. Therapeutic doses of oral anticoagulants and of imipramine for 6 months each did not arrest disease progress. In 1987 signs of generalized dementia were noted. The patient became unable to find his way around the city or within his house, could not perform shopping or banking tasks or be left on his own.

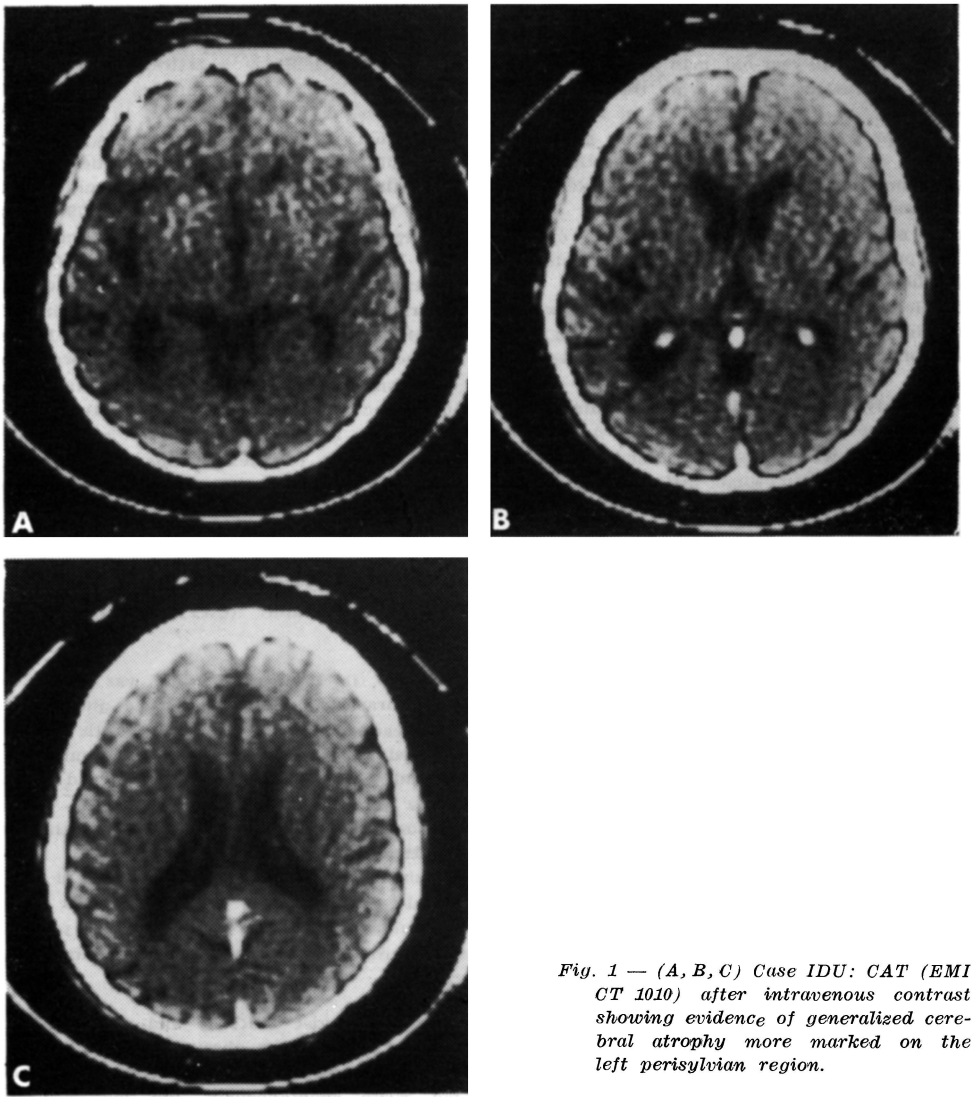

Fig. 1 — (A, B, C) Case IDU: CAT (EMI CT 1010) after intravenous contrast showing evidence of generalized cerebral atrophy more marked on the left perisylvian region.

COMMENTS

We have described a patient who over a period of 7 years evolved from a picture of focal left perisylvian dysfunction with difficulty handling written or verbal information to a more generalized language dysfunction, including transcortical motor aphasia, nominal aphasia, alexia, dysgraphia, dyscalculia and finally to generalized Alzheimer's dementia [3], except that even at this stage the speech and reading disorders were more pronounced than expected for classical Alzheimer's dementia [5]. This patient is similar to the 6 cases of Mesulam [6] and to the 2 cases of Chawluk et al.[1]. Positron emission tomography of the 2 latter patients demonstrated decreased glucose metabolism in the perisylvian region of the left hemisphere, supporting clinical, EEG and CAT evidence of a localized left perisylvian disease process. No evidence of systemic or other neurological disorder has been found in these 9 pacientes. CAT provided some clue on the form of generalized atrophy more marked posteriorly on the left hemisphere before the onset of dementia. EEG has shown a focal disturbance of the left perisylvian region super-imposed on a generalized disturbance, the occurrence of this association

possibly depending on the length of disease evolution. None of the 8 previously desdribed cases had a family history of Alzheimer's dementia. These 9 patients differ from other previously described aphasias due to focal lesions because of the striking progress of the language deficit over a period of a few years [1,4,6,8]. Alzheimer's disease is usually characterized by a progressive dementia with evidence of generalized cortical dysfunction such as temporal and spacial disorientation, difficulty in recognizing familiar objects and persons, deficits of recent memory, agnosia and ataxia [2]. Language disturbances in Alzheimer's disease are usually considered to be a complement of the progressive generalized dementing process [2]. Mesulam [6] and Chawluk et al.[1] emphasized the progressive nature and severity of the speech disorder with relatively little deterioration of personality or other cognitive and intellectual functions in their patients. Of their 6 initial patients 5 showed anomic aphasia and 1 had pure word deafness at presentation. EEG showed left hemisphere slowing and CAT indicated left hemisphere atrophy in at least 5 of the 6 patients. In one patient auditory evoked responses showed evidence of dysfunction of the left auditory area. The language deficit progressed in severity and involved other language functions as running speech, auditory repetition, writing and comprehension of written texts at more advanced stages of disease. The most common non-speech related deficit observed in their patients was acalculia. Two of the 6 patients developed a generalized dementia 7 years after onset of the aphasic disorder [6].

Although it may be considered that Alzheimer's disease encompasses a spectrum of clinical presentations ranging from focal signs such as aphasia to evidence of generalized dementia starting with the classical deficit of recent memory [2,9], we tend to support the view of Mesulam [6] and Chawluk et al.[1] that slowly progressive aphasia without generalized dementia should be considered a separate syndrome. Their cases as well as our own illustrate the striking difference on clinical presentation from the usual cases of Alzheimer's dementia [3,5] and from cases where aphasia preceeds dementia by shorter periods [9]. As the etiology of Alzheimer's and related disorders is obscure and diagnosis is primarily based on clinical history and examination, a pragmatic approach dictates that these syndromes should be kept separate until the issue is elucidated by further reports and hopefully by formal studies.

The diagnosis of slowly progressive aphasia is made on the basis of clinical history and examination and on complete medical and neurological evaluation showing no evidence of other disease processes. The prognosis on the 9 cases reported has been bleak with steady progression to complete aphasia and to generalized Alzheimer's-like dementia in 3 cases followed for approximately 7 years. In 6 other cases the aphasia progressed but follow-up has not been long enough to allow a conclusion about the onset of dementia. There is no specific therapy and in our case there was no response to specialized speech therapy.

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