PICK'S DISEASE IN OLD AGE

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PICK'S disease most commonly begins in the midlife period. Sjogren, Sjogren and Lindgren (1952) in their series of 18 anatomically verified cases of Pick's disease found that the average age of onset was $54 \cdot 5 \pm 2 \cdot 7$ years. Escourolle (1956) segregated from the literature 184 cases in which he considered the diagnosis of Pick's disease to be "indisputable" and demonstrated therefrom that the sixth decade is the period of highest incidence. Pick's disease, however, has frequently been reported to erupt in the seventh decade and occasionally in the eighth, ninth and tenth decades. (Bonfiglio, 1926; von Braunmühl, 1928; Moyano, 1932; Sjogren, Sjogren and Lindgren, 1952.)

Since Pick (1892) first studied cerebral atrophies with the single aim of clarifying contemporary concepts of aphasia, there has been considerable difference of opinion on what constitutes the disease which now bears his name, but the following brief definition may be said to accord with modern application of this term. The anatomical basis of Pick's disease consists of a circumscribed cerebral atrophy, the sites of predilection being the frontal and temporal lobes. Such atrophy involves both white and grey matter and is associated with disappearance and distortion of ganglion cells in the atrophic gyri. "Ballooning" of nerve cells and the appearance of silver staining inclusions in diseased cells are frequently but not invariably observed. (Escourolle.) The profusion of neurofibrillary tangles and senile plaques which constitutes the characteristic histological findings in senile atrophy and Alzheimer's disease is notably absent. It is generally agreed, however, that when Pick's disease occurs in aged patients, the presence of a few senile plaques does not invalidate the diagnosis.

The clinical picture described in reported cases of Pick's disease has in the past lacked uniformity but considerable agreement on this subject has been reached in recent work. (Delay, Brion and Escourolle, 1957; Robertson, le Roux and Brown, 1958). Delay *et al.* in their catalogue of the features which serve to differentiate the dementia of Pick's disease from that of Alzheimer's disease, stress the relative retention of memory and spatial orientation in the early stages of Pick's disease. They describe the mood as varying from euphoria to apathy and emphasize the absence of delusions and hallucinations. They instance the progressive amnestic aphasia, the iterative phenomena, the striking diminution in attention, the defects of judgment and the total lack of insight. Robertson *et al.* from a series of three cases of Pick's disease studied in this hospital, came independently to similar conclusions. They consider that the most striking feature of the progressive intellectual deterioration seen in Pick's disease is the absence of many of the spatial *and* temporal defects enumerated in lesions of the parietal lobe (Critchley, 1953). They attribute this finding to the relative integrity of the parietal lobe at autopsy. In their list of personality traits Robertson *et al.* include egocentricity, stereotypy, perseveration and restlessness, an absence of self-criticism and self-restraint, a placid or euphoric mood and a marked disinclination to co-operate in formal testing.

Reported Cases of Late Onset

Five cases of Pick's disease with an onset after the age of 70 have been reported, but only three of these are supported by adequate data. The case of Bonfiglio (1926) relating to a woman of 80 lacks post-mortem verification. No histological report accompanies the case of Moyano (1932). He describes a female patient whose symptoms erupted ten months before her death at the age of 92, and makes the diagnosis of Pick's disease on the basis of temporal lobe atrophy found at autopsy. Escourolle eliminates these cases from his list of "indisputable" cases of Pick's disease. von Braunmühl (1928) reports the case of a man of 81, admitted to hospital immediately after a street accident, with a history of progressive intellectual deterioration extending back over the previous two years. Autopsy performed eight months later revealed fronto-temporal atrophy and also an old subdural haematoma overlying the right hemisphere. Histological examination demonstrated "ballooned" cells, silver inclusions and senile plaques, but the latter were few in number. This intracranial complication of subdural haematoma in von Braunmühl's case has also been reported by Escourolle (two cases) and neither author considers that the presence of a subdural haematoma invalidates the diagnosis of Pick's disease. Sjögren, Sjögren and Lindgren in their series of 18 histopathologically verified cases include two female cases of late onset with predominantly frontal atrophy. One patient developed symptoms at the age of 72 and died five years later, while the other's illness commenced at the age of 74 and was of two year's duration.

Reported hereunder is a case of Pick's disease of late onset (at the age of 72), long duration (17 years) and relatively benign course.

Case

F.T.M., aet 80, formerly a shorthand writer to the Law Courts, was admitted to the Royal Edinburgh Hospital (Craig House section) on 9th December, 1952. He had previously been resident in a private psychiatric hospital for five months.

Family and Personal History

His father, paternal grandfather and great grandfather all lived to an advanced age without at any time exhibiting mental symptoms. His mother died of an unspecified physical illness in middle life. One spinster sister was described as having been "slightly eccentric" and another had a nervous illness in middle life from which she made a complete recovery. Otherwise there was no history of mental or neurological disease in the direct or collateral lines.

All available evidence indicated that the patient had been a highly intelligent man. After qualifying in law, he was offered and accepted the post of official shorthand writer to the courts. He was profoundly interested in Celtic literature and in the genealogy of Scottish families and had published a book and articles on these subjects which earned him the fellowship of a learned society.

History of Illness

The history was obtained from his two sons, his wife having died four years before his admission to hospital. Neither son had been in daily contact with the father, and therefore their accounts tended to be fragmentary. The patient retired from active court work in 1937, but wartime depletion of staff resulted in his occasional recall to the courts in the early war years. He continued to administer his firm of shorthand writers till 1946, when one of his sons, returning

home from war service, found him quite unfit to carry this responsibility and arranged the transfer of his firm to other hands. Both sons dated the onset of symptoms to 1944, when the patient was observed by his wife to have difficulty in reading newspaper headlines. Synchronously, he appeared to suffer from "loss of memory", but the informants were unable to quote specific incidents to illustrate this loss. Two years later his behaviour became abnormal. He had always been preoccupied with the rights of pedestrians, but now this trait became accentuated in a bizarre fashion. He was observed to walk backwards across the city's main street in front of oncoming traffic, to insert himself between moving trams and to step in front of buses. His habits showed progressive deterioration, and while travelling in public vehicles he would spit on the floor or out of the window and become abusive when remonstrance was made. He frequently expressed the belief that other people were mad. He had no insight into his condition. After his wife's death in 1948, a housekeeper was engaged to look after him, and later he went to live in a hotel, whence he was transferred to a private mental hospital in 1952. He had now become a constant embarrassment to the police, since he still travelled round the town, holding up buses which he wished to board at any point on their route, seemingly oblivious of the regulation stopping places.

Physical Examination

The patient was a healthy-looking old man of small stature. He moved with a clumsy, shuffling gait. There was no evidence of peripheral or fundal arteriosclerosis. B.P. 110/64. Examination of the C.N.S. revealed no abnormality. A large right scrotal hernia was present.

Mental Examination

The patient was somewhat untidy and slovenly in his attire and was in the habit of hoarding food in his pockets and burning holes in his clothing with his cigarettes. He appeared most contented when allowed to lie in bed, peacefully smoking. When bed was proscribed, he sat in an armchair near the fire with his feet up, smoking continuously. He hoarded cigarette ends, out of which he manufactured new cigarettes. At times, he sang boisterously, utilizing instead of words the doh-ray-me scale, e.g., doh-me-fah-fah-ray-doh, doh-soh-soh. Or he might more quietly hum a tune. His melodies were usually recognizable. He was able to attend to his toilet and feed himself.

During interview he was abrupt in manner. His attention was difficult to hold, and he made no attempt to persevere if he could not immediately comprehend or fulfil what was asked of him. He admitted his inability, however, and never confabulated. His flow of conversation was normal when allowance was made for a reduction in vocabulary and simplification of syntax. When he forgot a word he would endeavour to convey its meaning by circumlocution or gesture. His talk was liberally interspersed with certain stereotyped phrases, e.g., "I'm just a daft old man", "I'm just a funny old man". "My brain's all to pot". "I don't know anything". He co-operated poorly in routine tests. He refused to name objects presented to him, saying, "I don't know anything or anybody, I'm just a funny old man". Shown a watch, he read the time accurately, but was unable to name the object he was looking at. He was observed to read his newspaper diligently every day, but when questioned, invariably declared "I can't understand a word of it". His memory when assessed on his answers to questions appeared grossly defective, but it seemed doubtful whether this was a valid estimation in view of his speech defect, his inattention and unwillingness to concentrate. He spontaneously volunteered

some incidents from the past at the beginning of an interview. Observation of his behaviour in the ward revealed that he was quick to notice all that was going on around him and, in particular, any change in the programme of his daily activities, for example, the non-arrival of his morning paper. He remembered faces, although failing to name the individual persons. He was unable to give the day, month or year. Nevertheless, he was able to compute the passage of time, since he always returned from his daily walk at the appointed hour. He never lost his way in a ward block with many ancillary rooms and offices. Every day found him walking alone in the extensive hospital grounds, and he never failed to find his way back to his ward.

Subsequent Course

Throughout the illness his affect remained one of euphoria. A gradually progressive deterioration ensued which was especially noticeable in his personal habits. Increasingly frequent spitting and later urinating in inappropriate places necessitated the cancellation of his parole and in May 1955 his move to a ward for more deteriorated patients. The satiation of his appetite now became his *raison d'être*, and he would ask or begin his search for food within minutes of finishing a meal. At other times he sat quietly for long periods keenly watching the movements of those about him, and he utilized his observations to choose an appropriate moment for stealing food from other patients or for closeting himself alone in the ward kitchen. Nevertheless, he frequently and earnestly insisted that he had not eaten for some years!

In June, 1958, his general enfeeblement was such that he was transferred to a geriatric ward. Here he believed he was in a hotel and referred to the ward sister as "the lady of the house". He endeavoured to obey her instructions implicitly and became irritable if thwarted by others. He occasionally talked to her about the peculiarities of the other "guests". He was well-mannered, never aggressive, appreciative of personal service and a favourite with the staff. He still smoked and always made a point of extinguishing his cigarette ends. With discouragement, his spitting habit became less frequent, but his tendency to steal food persisted. Sometimes incontinent, he always appeared apologetic on these infrequent occasions. He was able to participate in a simple game involving catching and throwing a ball. He was upset by any deviation from his usual daily routine and several days would elapse before he could accustom himself to a change (e.g., of bed position). He checked the time frequently throughout the day and knew when it was time to go to bed. His tolerance of drugs was low and he became ataxic after only twenty grains of chloral hydrate and on one occasion after twenty-five mgs. chlorpromazine.

His speech continued to show correct syntax, although his vocabulary was meagre. He still sang loudly and his melodies remained recognizable. He was frequently seen standing at the window praying aloud "Dear Lord, make me a good man". Latterly, he was seen to look up at the sky and say "I'm coming dear, I won't be long", which on questioning he said was directed to "my dear wife up there". He rarely initiated conversation and whether or not he replied to questions seemed to depend on the enquirer's identity. He invariably answered when questioned by the ward sister, but might or might not respond if addressed by a member of the medical staff.

He was able to recognize an old photograph of himself in which he was dressed in the costume of Sir Walter Scott. His former interest in the newspaper lessened, but he continued to read parts aloud and made appropriate comments, e.g., of an accident, "how terrible"; or of a murder "how wicked". He could

never discuss an article and would plead "I don't understand, I'm just an ignorant old man". The shuffling gait noted at admission persisted unaltered.

His general deterioration had been accelerated by respiratory infections in 1958 and 1959, but it was only in his last months that he had to be assisted with feeding, washing and dressing. His speech was now almost entirely emotive; food searching and noisy spells were much less frequent and he spent the greater part of the day in bed. Towards the end of February, 1961, he had a small haematemesis, subsequently developed bronchopneumonia and died a week later on 2nd March, 1961, aged 89.

Necropsy Findings (only positive findings charted)

The *lungs* shows patchy basal congestion with early bronchopneumonic consolidation. Oesophagus, stomach and duodenum were normal, and the source of the minor haematemesis was presumed to be a gastric erosion, now healed.

The *brain* weighed 1,300 gm. The main arteries showed mild atheroma. The leptomeninges showed a little diffuse thickening, most marked over the cerebral convexities. There was mild generalized symmetrical atrophy of the cerebrum. Accentuation of this atrophy was apparent in both frontal lobes while there was gross symmetrical wasting of the temporal lobes. The areas most affected were the temporal poles and the anterior portions of the lobes. The posterior half of the superior temporal gyrus and the posterior third of the middle and inferior gyri were relatively normal.

Serial coronal slices of the cerebrum confirmed the generalized atrophy and its severe local accentuation in the temporal lobes. The lateral and third ventricles were dilated and the aqueduct normal. The brain stem and cerebellum were normal externally and in section. The fourth ventricle was normal.

Microscopic Observations

Sections of the temporal lobes showed thinning of the cortex with a profound loss of ganglion cells and an astrocytic gliosis, the superior, middle and inferior gyri being affected in that order of increasing severity. The white matter showed extensive pallor of myelin staining, with perivascular shrinkage which amounted in places to the formation of small cysts with a few pigmented histiocytes in their walls. Silver stains failed to demonstrate argyrophil inclusions, tangles or senile plaques. The frontal lobes showed a less severe outfall of ganglion cells with gliosis and a few scattered senile plaques. No neurofibrillary tangles were seen. The small arteries were normal.

Analysis of the Clinical Features

The patient was not seen by the present writers till three years after his admission to hospital and we accepted at that time the initial diagnosis of senile dementia, since his advanced age and the long duration of his illness seemed to eliminate any alternative diagnosis. After the autopsy findings were known, renewed scrutiny of the case notes combined with a few additional details from the patient's two sons served to build up a clinical picture characteristic of Pick's disease. The salient features are tabulated hereunder.

Spatial and Temporal Orientation

The first signs of intellectual deficit and personality changes were observed eight years before the patient's admission to hospital. Throughout these eight years he was in the habit of journeying unaccompanied through a busy city, without mishap to himself, albeit his foolhardy behaviour aroused consternation

in observers. Once in hospital, he quickly orientated himself to a hospital ward, to its ancillary offices and to the extensive hospital grounds. He was able to dress and feed himself, wielding knife and fork with conventional skill, and his performance was not impaired until a few months before his death. He could read the time on a clock face. His frequent consultation of the ward clock and his regular return from his daily walk at the appointed time argued an awareness of the passage of time. The careful attention to detail and timing which he showed in his plots to steal food indicated his retention of a concept of order and sequence, which in itself is linked to a time sense.

Nominal Aphasia

The patient's spontaneous speech showed a progressive reduction in vocabularly with preservation of normal syntax. Dysarthria and paraphasia were never observed. Perseveration was an outstanding feature, exemplified by his monotonous reiteration of stereotyped phrases, e.g., "I'm just a daft old man", "I'm just a funny old man". "My brain's all to pot". Systematic tests for aphasia could not be applied because of his attitude of negation and inattention. He rarely succeeded in naming objects, but could indicate their use by circumlocution or gesture. His pertinent comments on the headline news in his newspaper proved that he was able to read and understand what he had read, but his comprehension was limited to the simple and concrete. He invariably refused requests to read aloud words or sentences.

His singing of familiar songs indicated his retention of a sense of melody. (Robertson *et al.* report a similar finding in two of their cases). He continued to converse and make himself understood, despite a now grossly impoverished vocabulary until his brief fatal respiratory infection. This last must be counted atypical, since Pick's disease commonly concludes in a prolonged "terminal stage of mutism and hebetude" (Schneider, 1929).

Memory

Memory defects must inevitably form part of any dementing process, but the slow impairment of memory in Pick's disease, first emphasized by Kahn and Thompson (1934) has since been corroborated by many authors. Memory function must of necessity be assessed on the patient's behaviour, since the amnestic aphasia and characteristic inattention render dubious the results obtained from verbal tests. Observation of our patient revealed that he remembered faces (although unable to name individuals) and events of personal significance, e.g., he expressed annoyance when his morning paper did not arrive. He rarely failed to carry out instructions which involved the continuous recollection of several details. He was aware of the significance of death, remembered his religious instruction, and anticipated a reunion with his wife, who, he recollected, had died several years before. Already tabulated in the first section are examples of the patient's ability to orientate himself in space and time, some of which could have been appropriately described under "memory".

Personality Changes

Egocentricity and a blunting of social judgment, illustrated in the patient's bizarre behaviour in traffic and his lack of concern for other road users, were evident at an early stage. Following admission, similar traits were exemplified in his habit of stealing food from other patients. A euphoric mood was observed throughout, as was also a marked disinclination to co-operate in mental tests (which last implies a total absence of catastrophic reaction). Perseveration and stereotypy, reported by Robertson *et al.*, as characterizing the behaviour of their

patients during their independent pre-hospital existence, could not be elicited in this case, where details of the earlier stage of the illness were unfortunately meagre. These traits are unlikely to be obvious once the patient is in hospital since institutional life confers its own kind of stereotypy.

Physical Symptoms

The patient's clumsy shuffling gait was the only physical abnormality noted. Anomalies of gait have frequently been reported in Pick's disease (Caron, 1934). The hyperalgesia found by Robertson *et al.*, in all their cases and considered by them an important diagnostic sign was not elicited in this case but this does not exclude its presence at an earlier period. Robertson *et al.* found that this hyperalgesia was transient and notably absent in the later stages of the illness.

Pathological Findings

Superimposed on a generalized mild cerebral atrophy—an expected finding in a patient of this advanced age (89)—were a severe symmetrical temporal lobe atrophy and a frontal lobe atrophy of minor degree. The salient histological feature was an outfall of ganglion cells from the atrophic gyri. No neurofibrillary tangles were seen. A few scattered argyrophil plaques were present in frontal lobe sections, compatible with the patient's age. These were notably absent in the temporal lobes, the areas of maximum atrophy.

Duration and Course

The duration of the illness in this case was unusually long—17 years, but this has been exceeded by a year in a case described by Schenk (1951). Equally unusual was its relatively benign course—our patient continued to show interest in his surroundings, to maintain verbal and emotional rapport with the nursing staff and to remain ambulant until a few weeks before his death.

Summary

A description is given of a patient who developed Pick's disease at the age of 72, and died aged 89. This case along with others of late onset, indicates that this so-called "pre-senile dementia" must also be included in the differential diagnosis of organic psychoses in the senium.

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