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Limbic dementia[1]

GENEROSO G. GASCON AND FLOYD GILLES

From the Departments of Neurology and Neuropathology, Harvard Medical School, and the Children's Hospital-Peter Bent Brigham-Beth Israel Neurology Residency Program, Boston, Massachusetts, U.S.A.

SUMMARY This is the second reported case, known to the authors, of complete, but selective, limbic lobe destruction in previously normally functioning central nervous systems. Both cases had an amnestic syndrome, whose characteristics were essentially similar to amnestic syndromes previously documented with less complete limbic destruction, with one difference—confabulation remained a prominent feature in the chronic stages of the memory disorder. Our patient exhibited a behavioural syndrome similar to that reported by Klüver and Bucy in monkeys with bilateral anterior temporal lobectomies. Klüver-Bucy like syndromes in man have usually been reported with surgical lesions, usually in patients with pre-existent brain disorder. Our case illustrates that the syndrome can be produced by necrotizing encephalitic lesions. We suggest that the combination of the above two syndromes is essentially a 'limbic dementia'.

Permanent arrest of recent memory functions has been related to bilateral hippocampal damage (Scoville and Milner, 1957; Penfield and Milner, 1958; Victor, Angevine, Mancall, and Miller-Fisher, 1961; Whitty, 1962; Drachman and Arbit, 1966; DeJong, Itabashi, and Olson, 1969; Pribram, 1969). In the longer-known amnestic syndrome of Korsakoff's psychosis, loss of mamillary bodies (Brion, 1969) and damage of medial dorsal nuclei (Victor, 1964) have been implicated as the critical anatomical deficits. These and other limbic structures (Angevine, Locke, and Yakovlev, 1964) are also implicated as the anatomical substrates of emotion (Papez, 1937; MacLean, 1949, 1952).

The suggestion that a demented condition could be an expression of damage to limbic structures has been made previously (Corsellis, Goldberg, and Norton, 1968). The clinical findings in our case add to the spectrum of chronic behavioural and intellectual changes which may result from extensive limbic and paralimbic destruction.

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CASE REPORT

G.M., a 26 year old right-handed mother of two, was admitted to the Peter Bent Brigham Hospital for the third time on 31 October 1967 in an acute confusional state.

She had graduated from high school at the age of 16, had a satisfactory employment history, and was described by her husband as a self-sufficient housewife. Hodgkin's disease was first diagnosed in July 1964 by axillary lymph node biopsy. She was seen by a psychiatrist on 28 August 1964 because of emotional lability manifested by periods of hypomanic behaviour alternating with crying. She demonstrated features of an acute anxiety reaction in a basically healthy personality. The reaction was transient and repeated neurological examinations during the previous two admissions were within normal limits.

On 24 October 1967 she developed headache, fever, nausea, and vomiting. These symptoms subsided, but were followed on 30 October by confusion, inappropriate affect, antagonistic attitudes, and agitation. Over the next day she lapsed into somnolence and was readmitted with a temperature of 38·9° C (102° F).

NEUROLOGICAL EXAMINATION She was distant, hostile, oriented only to person, and unable to remember recent or remote events. She could subtract serial 7s only to 93, and proverb interpretation was deficient. The rest of the examination was unremark-

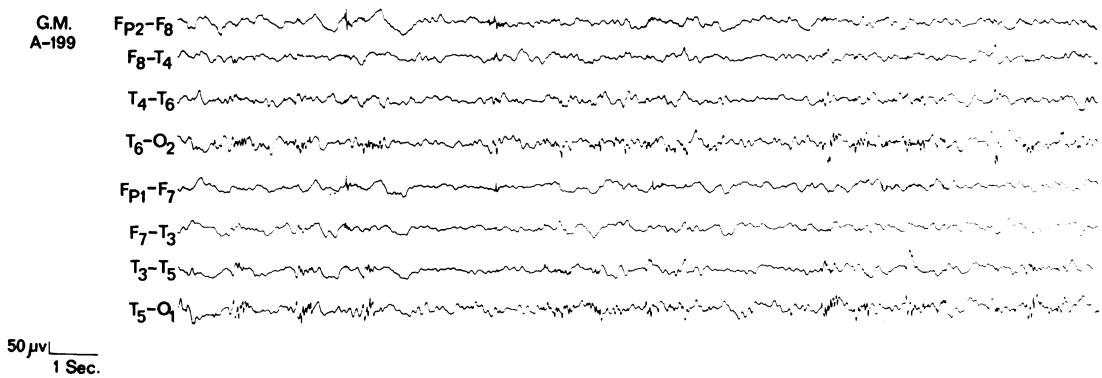

FIG. 1. *EEG on 10th day of illness. Bilaterally there are spike foci in posterior temporal regions. Background activity is generally slowed.*

able. Cerebrospinal fluid examination on 1 November 1967 showed 11,648 erythrocytes per c. mm, no leucocytes, a sugar concentration of 54 mg/100 ml., and protein concentration of 28 mg/100 ml., with slightly positive Pandy test.

From 1 to 10 November 1967 the level of consciousness waxed and waned. No elementary neurological signs were seen, except for an abnormal left plantar response, at a time when there was evidence of midline shift to the left by A-scan echoencephalography (4–6 mm) and right internal carotid arteriography. The latter also revealed elevation of the right middle cerebral artery and stretching of the anterior temporal artery. Electroencephalograms first showed bilateral widespread delta background activity, then bilateral posterior temporal periodic spike foci (Fig. 1). A pneumoencephalogram revealed a dilated right temporal horn.

From 13 November to 4 December, repeated bedside testing showed that she spoke fluently and spontaneously in short, grammatically correct, sentences, and showed no gross motor or coordination deficits. She was oriented only to person. The place was 'your apartment' or 'here' or 'a restaurant', and the date was 'last Sunday'. She was unsure about the number of her children. She did not remember the examiner's face or name after three minutes' absence. She had excellent comprehension of speech, but found it difficult to name objects. A thermometer was 'a needle' and a watch was 'a clock'. In her conversation the same themes frequently returned, and she often confabulated. She

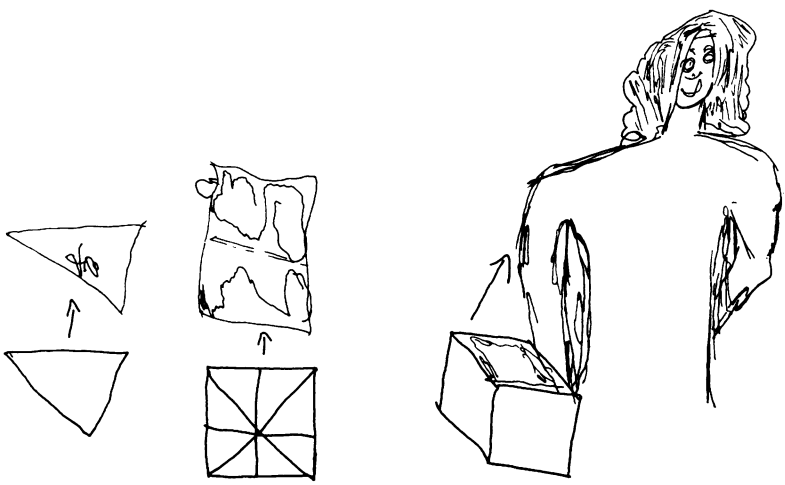

FIG. 2. *Examiner's drawings of common geometrical figures are at the bottom. Arrows point to G.M.'s copies. Of greatest interest is her copy of the cube. Her initial attempts are seen within the top surface. Continuity with the external human figure is apparent.*

smiled automatically in response to the examiner's smiles, but without affective participation.

She named colours correctly. She could repeat the names of three objects immediately, but after a minute could not spontaneously recall them or recognize them when given multiple choices. She exhibited a marked retrograde amnesia, with islands of preservation. The furthest point was difficult to define but seemed to extend back at least four years, for she did not seem to be aware of President Kennedy's assassination. She had an anterograde amnesia for events beginning with her hospitalization. She correctly read aloud a paragraph on the Russian educational system, but immediately afterwards described what she had read as 'school'. After looking at a two-page spread of photographs for one minute, she could not tell the examiner the relative locations of various pictures. She wrote her name, address, and a sentence describing the weather correctly and legibly with her right hand She copied geometrical figures in a bizarre fashion. She drew a triangle with aimless graphic perseveration, and, with a square, she showed the 'closing in' phenomenon. She drew a girl when asked to copy a cube (Fig. 2). She denied that anything was wrong with her health or memory. Proverb interpretation was meaningful.

Formal psychological testing could not be completed fully because of impaired attention span and regressive behaviour manifested by alternate negativism and petulance. However, there were distortions in the few Bender-Gestalt figures which she attempted to reproduce from memory after a five second viewing. She could count backwards from 20 in 17 seconds with one error (omitted 11). Counting by 3's and starting at 1, she reached 22 after 90 seconds. She recalled six digits forward, but could not reverse them. With the Rorschach test she went through only seven cards, taking seven minutes. Perseveration occurred starting with card IV.

Over the next two weeks, an additional behavioural aberration was observed. She frequently and indiscriminately put food and objects, even faeces on one occasion, into her mouth. She was also frequently found searching aimlessly through other patients' dressers. When eating she often used fingers instead of utensils.

Formal speech and language testing (Table) on 7

TABLE

SUMMARY OF SPEECH AND LANGUAGE EVALUATION ON G.M., SIX WEEKS AFTER ACUTE CONFUSIONAL STATE

Spontaneous speech: fluent without dysarthria
Peabody Picture Vocabulary Test: consistent with educational background
Auditory comprehension: intact for short oral questions, deteriorated on long paragraphs read aloud to her correctly matched pictures, colours, forms, numbers, letters, words, and sentences to oral verbal stimuli pointed to named body parts identified all presented non-verbal sounds
Visual comprehension: matched printed words to pictorial counterpart
Repetition: correctly repeated all numbers, words, and sentences
Serial speech: correctly counted to 20, recited days of week, months of years, alphabet
Naming: correctly named body parts and pictures of common objects
Reading: correctly read aloud two passages of moderate difficulty, though could not answer questions about them unable to answer questions related to paragraphs of greater than three sentences which she had read silently
Writing: no impairment, including spelling and punctuation
Arithmetic: no gross deficits. Could tell time, set a model clock, do complicated reasoning problems and simple arithmetic
Drawing: drew circle, square, and cross on command. Drew a triangle when asked to draw a diamond

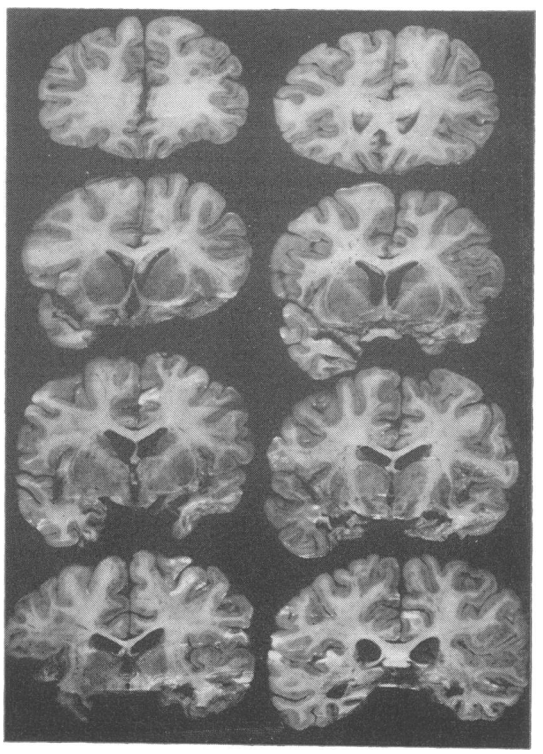

FIG. 3. *Posterior surface of coronal slabs of brain. There is marked destruction of posterior orbital, insular, and anterior and medial temporal lobes, including amygdalae. The inferior and lateral surfaces of the left temporal lobe (left lower slab) were utilized for viral cultures.*

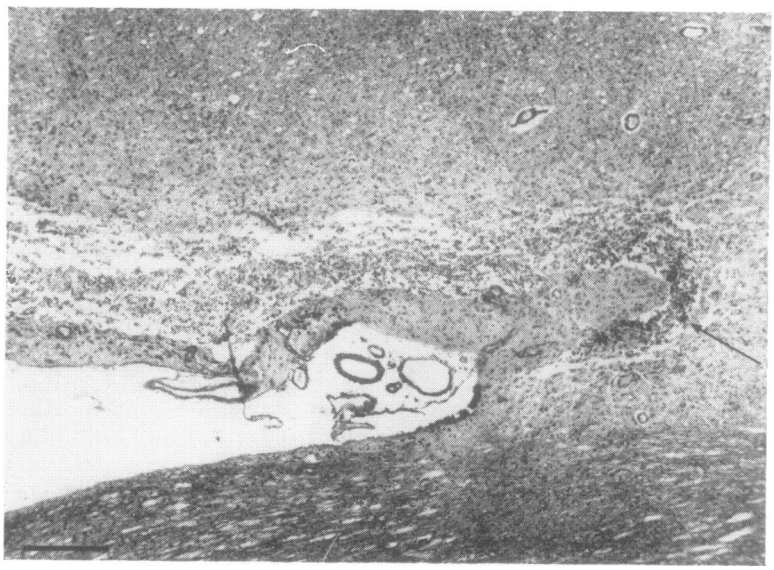

FIG. 4. (*H and E. Marker = 0·5 mm*). *Gyrus cinguli above, corpus callosum below. Laminar central total cystic necrosis is surrounded by a layer of macrophages and astrocytes. Mineralized neural debris (arrow) is located at right.*

FIG. 5. (*Luxol fast blue-Cresyl violet. Marker = 0·25 cm*). *There is total necrosis of hippocampal (small arrow), parahippocampal (large arrow), and fusiform gyri.*

December revealed, in addition to previous clinical bedside testing, frequent circumlocutions and illogical train of thought. She had exhibited bizarre naming before the formal testing, but now she correctly named all body parts and pictures of common objects. She was also able to name subordinates to class words; for example, 'car' then 'Cadillac'.

Subsequently, there was no essential change in her mental status. Neither the retrograde nor anterograde amnesia improved. She continued to show marked hyperphagia, much wandering around the ward, and inappropriate jocularity.

She sustained a right retinal detachment, further increase of Hodgkin's infiltrate in the lungs, and lung abscess, and died in February 1968.

After death the following viral antibody titres were received.

Cytomegalovirus antibody titres:

9 January 1967	1:16
10 April 1967	1:16
11 December 1967	1:256

Herpes simplex:

9 January 1967	1:256
11 December 1967	1:4096

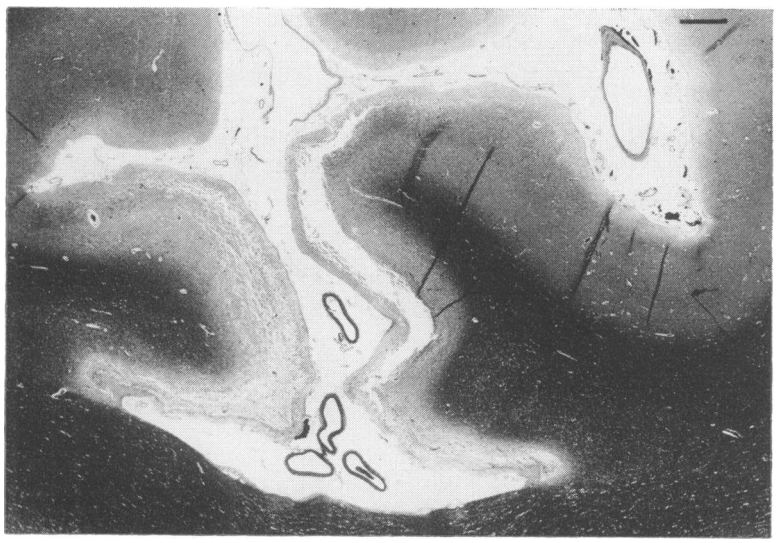

FIG. 6. (*PTAH. Marker = 0·1 cm*). *The gyri cinguli are necrotic bilaterally. The corpus callosum lies below.*

NECROPSY FINDINGS The general necropsy findings included residual Hodgkin's granuloma in lung, spleen, and vertebral bodies as well as bronchopneumonia. She had a severe depletion of bone marrow and splenic lymphoid tissue. Fresh left temporal lobe tissue obtained at necropsy was inoculated into Wi-38, human embryonic kidney and monkey kidney media. All cultures were negative.

The brain was fixed in 15% unbuffered formalin, slabbed, blocked, and processed by standard methods and 15 μ sections were stained by haematoxylin and eosin or Luxol Fast Blue with Cresyl Violet.

Widespread destruction of limbic and paralimbic structures bilaterally was characterized, for the most part, by its symmetry, with the general exceptions that temporal and insular isocortex was somewhat more extensively damaged on the right. In most regions the destructive process included juxta-allocortex and isocortex adjacent to damaged limbic structures (Fig. 3).

The regions of damage were characteristically cystic centrally and bordered by a layer of hypertrophic astrocytes and astroglial fibrils. Large numbers of foamy macrophages were present centrally, as well as in the border, and were particularly prominent around blood vessels where they widely distended the perivascular space (Fig. 4). Frequently, astrocytic nuclei were large and bizarre and, while often containing nuclear bodies (Krishan, Uzman, and Hedley-Whyte, 1967), no intranuclear inclusions could be identified. An inflammatory response was, at best, minimal. It usually consisted of several lymphocytes lying around a capillary or a venule and an occasional plasma cell. The latter were frequently bizarre and contained multiple nuclei. The process ended abruptly at its borders and frequently within the space of one or two microscopic fields normal cortex was encountered. In spite of a diligent search no intranuclear inclusion bodies were identified. In the border of these lesions small foci of mineralized neurones or glial cells were sometimes encountered. No inflammatory response was encountered in small necroses at the depths of sulci in the lateral, temporal, or occipital lobes.

The bilateral damage of the anterior perforated space and substantia innominata extended into the septal cortex medially, the ventral portion of the nucleus accumbens septi superiorly, the posterior orbital isocortex anteriorly, as well as the inferior insular isocortex laterally, the latter more so on the right. Several preserved portions of the nucleus basalis were bilaterally present posteriorly and medially. Insular damage on the right extended into the isocortex of the superior tip of the Sylvian fissure, on the inferior surface of the right third frontal gyrus in its middle third, and deep into the extreme capsule and peripheral parts of the right claustrum.

The entire hippocampal formation as well as the parahippocampal gyri bilaterally had been destroyed (Fig. 5). The destructive process included the pyriform cortex and subjacent amygdalae bilaterally. All of the adjacent juxta-allocortex and the adjacent isocortex of the temporal lobe were involved. The process had extended to the fusiform gyrus on the left, and the fusiform and third temporal gyri on the right anteriorly. The temporal tips had been destroyed

bilaterally. The anterior commissure had undergone severe degeneration and was gliotic. The right fornix was markedly atrophic; the left much less so. Both mamillary bodies were intact; the right was moderately atrophic.

The gyrus cinguli was partially destroyed anteriorly bilaterally (Fig. 6); the destruction extended from the subrostral gyri to the level of the splenium. The damage was more prominent on the right. The adjacent hippocampal rudiment was shrunken.

Isolated cortical necroses were present around the depths of sulci in the right occipital lobe medially and at the depths of the right first temporal sulcus.

The left thalamus was intact throughout. In the right thalamus there was a thin layer medially of total necrosis which extended from the ventral anterior nucleus anteriorly, through the medial aspect only of the dorso-medial nuclei to the medial portion of the pulvinar posteriorly. Lateral dorsal nuclei bilaterally appeared to be normal as did the intralaminar nuclei. No other lesions were found in the thalamus.

DISCUSSION

REPORTS OF INCOMPLETE LIMBIC DESTRUCTION Most human cases reported with behavioural or intellectual aberrations and correlated with lesions of the limbic system have shown, on post-mortem examination, incomplete involvement of limbic structures. We submit that this is because there are relatively few disease processes that would account for selective but complete involvement. The vascular supply is multiple, involving mostly branches of the posterior cerebral, anterior cerebral, and posterior communicating arteries, so that involvement of the major part of limbic structures by occlusive vascular disease is unlikely, though partial involvement has been reported (Glees and Griffith, 1952; Victor *et al.*, 1961; DeJong *et al.*, 1969; Faris, 1969). Neoplasm would have to be widespread to infiltrate the whole system, and, in fact, has not been reported, to our knowledge, although partial involvement is recognized (Malamud, 1967). Surgical lesions have been partial ones, such as cingulectomies and frontal lobotomies performed for schizophrenia, and with bilateral medial temporal lobectomies, which led to the recent intensive neuropsychological interest in memory (Scoville, 1954; Terzian and Dalle Ore, 1955; Scoville and Milner, 1957; Penfield and Milner, 1958). The congenital anomalies, such as arhinencephaly, nearly always end in early death but, if the infants survive, the clinical picture has usually been that of profound mental deficiency, although there have been rare exceptions (Nathan and Smith, 1950). There are no specific degenerative diseases of the limbic system as a whole. One possible exception is the report by Corsellis, Goldberg and Norton (1968) suggesting degeneration of limbic structures as a distant effect of carcinoma.

REPORTS OF COMPLETE LIMBIC DESTRUCTION In the past decade evidence has accumulated that an infectious disease, acute inclusion body encephalitis, an illness whose viral agent was unidentified in early cases, but shown to be *Herpes simplex* in later cases, often has a specific predilection for limbic structures (Rose and Symonds, 1960; Brierley, Corsellis, Hierons, and Nevin, 1960; Drachman and Adams, 1962). Only one report correlating extensive, but selective, pathological lesions largely limited to the limbic system with behavioural and mental changes is known to us (Friedman and Allen, 1969). We present the findings in this case of probable herpes simplex encephalitis as an addition to the spectrum of chronic behavioural and intellectual changes which may result from extensive limbic lobe destruction in man.

CLINICAL-PATHOLOGICAL ANALYSIS OF OUR PATIENT For purposes of discussion, we can divide the patient's chronic clinical neurological status into the behavioural aberrations and the memory deficit. A third area of concern, her naming difficulty, was prominent only in the acute stage of her illness, and was felt to be a non-aphasic misnaming syndrome secondary to the initial widespread cerebral dysfunction (Weinstein and Keller, 1964; Geschwind, 1967).

Behavioural aberrations These consisted of denial of illness, inappropriate jocularity, negativism, hyperactivity, short attention span, distractability, searching, and mouthing (hyperphagia and bulimea). Like patients with Korsakoff's syndrome and unlike most with bilateral surgical lesions of the hippocampal region, she was unaware of her memory deficits and denied any difficulty in remembering. In

fact, she denied that she was ill at all at a time when she was no longer acutely ill or obtunded, as in the syndrome described by Weinstein and Kahn (1955).

She was an amusing patient to talk to because the content of her speech was often inappropriately jocular, reminiscent of *Witzelsucht*.

Although she was apathetic shortly after the acute confusional state had ceased, she was unlike the patient with chronic Korsakoff's syndrome who usually exhibits lack of initiative and apathy, inertia, and indifference. She became quite hyperactive with much wandering about the ward, and exhibited motor restlessness such as constant swinging of her legs while sitting. Because she could not sustain attention and was so distractable, she was unable to sustain goal-directed activity. The quality of the patient's investigation of her surroundings—specifically the mouthing and oral tendencies—is not part of the usual syndrome of motor restlessness, hyperactivity, and distractability seen after encephalitis (Byers and Meyer, 1952).

Similar symptoms were reported in only one of the survivors of herpes simplex encephalitis seen by Drachman and Adams (1962). The cases of memory defect after encephalitis reported by Rose and Symonds (1960) showed hyperactivity, but no specific mention of global attentional disorder was made.

Many of the behavioural aberrations are similar to the syndrome resulting from bilateral ablation of the anterior temporal lobes, including uncus, amygdala, and hippocampus, in macaque monkeys, described by Klüver and Bucy (1939). Their monkeys exhibited: (1) 'psychic blindness', (2) strong oral tendencies, (3) a strong tendency to attend and react to every visual stimulus, (4) absence of the motor and vocal reactions generally associated with anger and fear, and (5) increased sexual activity. By 'psychic blindness' Klüver and Bucy meant that the animals responded indiscriminately to animate and inanimate objects and behaved as if all available objects belonged to one general class—something to be approached and examined. In multiple object tests, pulling-in tests, and random cage behaviour, the animals would pick up, mouth, and contact objects over and over again, as if they had not examined them before. At first this behaviour was equated with visual agnosia, but the difficulties with 'agnosia' as an explanation in a monkey have already been pointed out (Geschwind, 1965).

Our patient exhibited the first four of these characteristics. Although she was never observed to have overtly increased autosexual, heterosexual, or homosexual behaviour, a psychiatrist thought there was suggestive sexual connotation in her conversation, and that she exhibited confused courting of the examiner. Her responses on Rorschach testing to the first two cards were sexual associations.

Terzian and Dalle-Ore (1955) reported a 19 year old male, an epileptic with bilateral independent temporal foci, who underwent left, then right, anterior temporal lobectomy, including uncus and hippocampus, as a case of the Klüver-Bucy syndrome in man. The patient did not recognize anyone at first ('psychic blindness'). He showed near-catatonia for the first two weeks after operation, but then reacted to every stimulus. He had an insatiable appetite, and complete loss of any emotional behaviour, with indifference and amimia. Speech was normal, except that language was 'composed of very elementary sentences within the limits of his natural needs'. He had a serious disorder of present and past memory, which it was not possible to delineate fully. Up to this point their patient resembles ours. In addition, however, their patient showed a partial alexia and no spontaneous writing. His chronological sense of time was preserved, unlike ours. Also unlike ours, he had sexual exhibitionism, though no sexual aggression, and homosexual tendencies. He did not show oral tendencies or mouthing behaviour.

The patient of Friedman and Allen (1969) had repetitious motor activity, such as patting his knee and tapping his feet, and frequent impulsive behaviour, such as tactless, inappropriate remarks and mischievousness, though no specific mention of short attention span and distractability was made. The patient's mood was felt to be compatible with his premorbid personality, unlike our patient who underwent a definite personality change. Unlike our patient, their patient lost calculation ability, his sleep habits, thirst, and appetite were unchanged, and he lacked oral compulsions.

In addition to the fact that our patient

showed the behavioural components of the Klüver-Bucy monkeys, the bilateral anterior temporal lesions resemble those of the monkeys. We propose, therefore, that her behaviour can be characterized as an instance of the Klüver-Bucy syndrome in man.

Memory deficit Our patient showed a confabulatory-amnestic syndrome with anterograde and retrograde amnesia; the prototype of which is Korsakoff's psychosis (Talland, 1965).

Her deficit differed in that confabulation persisted as strongly in the chronic stages as it was in the acute stage, and remained easy to evoke (as with Friedman's and Allen's patient). With respect to memory difficulty, however, our patient and those with Korsakoff's syndrome are qualitatively similar.

Because of her short attention span and distractability, it was difficult to test her in a way that would prove conclusively to all observers that the inability to remember both verbal and non-verbal items was due to a specific deficit in memory processing, and not to the inability to concentrate. This criticism is especially relevant to those operational tasks which presented new material to learn.

However, it is difficult to accept an inability to concentrate as the entire explanation for defective responses, because even highly overlearned, familiar, and automatized items such as her birthdate, marriage date, and the number and names of her children could not be reproduced. Also, it would be difficult to account for her extensive retrograde amnesia, repeatedly documented by questions concerning past newsworthy events, such as her inability to recall one of the most emotionally traumatic events of the 1960s, President Kennedy's assassination.

Her responses to other cognitive testing stand in contrast to her defective responses on memory testing. She could not complete the WAIS, so that firm conclusions about her intellectual functioning, based on psychometric data, cannot be made. The main limitations that prevented completion of tests was attention span as well as negativism. However, to dismiss her as demented in the sense of marked loss of previous level of intellectual functioning is inaccurate. She could read, write, and do simple arithmetic. After the acute stage, she did not interpret proverbs concretely. The Peabody Picture Vocabulary Test, the one IQ test which she could complete, was consistent with her previous highest educational level (high school).

The features which clearly distinguish our patient from the Scoville-Milner group I patients (those with the most radical bilateral medial temporal lobe ablation) and those with Korsakoff's psychosis, are not the quality of the amnestic syndrome *per se*. Unlike both those groups, our patient exhibited the additional severe behavioural aberrations previously discussed. Moreover, although her lesions included those of the Scoville-Milner patients, she had little involvement of those areas said to be crucial for Korsakoff's psychosis—the mamillary bodies, or dorsomedial nucleus of thalamus.

Limbic dysfunction In a provocative article Yakovlev (1948) viewed behaviour as three different spheres of movement—the sphere of visceral motility; the sphere of motility of the outward expression of internal states, which includes the gamut of emotions (internal motions brought out which affect the animal or man but, *per se*, effect no change in the world of matter about it); and the sphere of motility of effectuation, which creates changes in the world of matter by use of parts of the body as tools. He postulated three anatomical systems which mediated the three spheres of behaviour. The entopallium mediates visceral motility, the mesopallium the emotions, and the ectopallium the sphere of motility of effectuation. The mesopallium extends from olfactory forebrain through the neuraxis and lies intermediate between the innermost system, the entopallium, and the outermost system the ectopallium. In the cerebral hemispheres the mesopallium includes the cingulate gyrus, retrosplenial and hippocampal gyri (collectively the limbic lobe of Broca), the orbito-mesial wall of the frontal lobe, and the island of Reil.

Papez (1937) proposed a theory that 'the hypothalamus, anterior thalamic nuclei, gyrus cinguli, the hippocampus and their connections constitute a harmonious mechanism which may elaborate the functions of central emotion, as well as participate in emotional expression'.

MacLean (1949, 1952) reviewed subsequent developments in neurophysiology which sup-

ported the Papez theory, with special implications for psychosomatic disease. The Klüver-Bucy monkeys were selected as most striking.

Based on our case and that of Friedman and Allen, the manifestations of complete limbic lobe destruction in previously normal brains seem to be an amnestic syndrome and a Klüver-Bucy-like syndrome. In the latter the most consistent feature seems to be, first, the emotional non-reactivity, manifested by lack of rage, and indifference, and second, a global attentional disorder manifested by short attention span, marked distractability, and impulsiveness. The presence of increased sexual expression and mouthing oral tendencies is inconsistent.

Teleologically speaking, complete limbic lobe destruction destroys a basic requisite for survival —the ability to detect and remember relevant (dangerous-safe) from irrelevant environmental stimuli. Subsequently, one of the manifestations of attempts to survive—anger or rage—disappear. Increased sexual expression may only indicate the reciprocal relationship between instinctive rage and sexual emotions—one ordinarily cannot have pleasurable sexual feelings while angry, and *vice-versa*.

Bilateral lesions of hippocampal region are sufficient, though not necessary, to produce memory disorder, since other lesions—for example, mamillary bodies—can also produce it. The added destruction of other limbic areas does not seem to alter the memory disorder except to make it more difficult to document because the general prerequisites for memory disorder, concentration and attention, are not intact.

Nevertheless, it was still possible to document that the limbic system is essential for transfer to long-term memory, or early consolidation, and also for retrieval of memories dating months to years back, though not essential for immediate, unconsolidated memory, which is probably mediated through the cerebral cortex. Although the limbic system is probably not necessary for long-term memories dating to early childhood, we were not able to document this in our patient.

The fact that the right temporal lobes were more completely destroyed than the left in both our case and that of Friedman and Allen raises the question of the effect of dominance on emotional behaviours. Our patient's affective behaviour quite closely fits that of right hemisphere-damaged patients with 'indifference reactions', consisting of jokes, mockeries, disinhibition, anosognosia, and minimization (Gainotti, 1972).

We cannot make specific clinical-anatomical correlations in our patient with regard to discrete behavioural aberrations, as, for example, relating decreased attention span and distractability to the bilateral hippocampal lesions as an example of the release of inhibitory functions of the hippocampus (Grastyán, 1959), or relating the lack of rage and anger to the bilateral amygdala lesions. She had too many other anatomical structures involved to make this kind of traditional clinical-pathological correlation. However, we propose that the structures involved do fall into a functionally interrelated unit, essentially that of Yakolev's mesopallium, Papez's and MacLean's visceral brain, or the limbic system. There was a right anterior temporal neocortical lesion (as in Friedman and Allen's case) and, although not strictly part of the limbic system, this area interconnects with more medial limbic structures such as amygdala and hippocampus. No significant permanent lesions were demonstrated in neocortical association areas.

We therefore suggest that this patient's syndrome was a 'limbic dementia', to emphasize that, although on superficial examination she may have appeared demented in the usual sense, the functions primarily lost were not those cognitive functions usually associated with 'intelligence', usually measured by psychometric tests, and often based on language, but those which imprint an affective quality, thereby giving meaning, to daily survival interactions with the environment and imprinting them into memory. Moreover, the neuropathological lesions were not those of the usual cortical dementias, but were confined essentially to hemispheral structures within the limbic system.

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