Pure psychic akinesia with bilateral lesions of basal ganglia

DOMINIQUE LAPLANE, MICHEL BAULAC, DANIEL WIDLÖCHER, BRUNO DUBOIS
Hôpital de la Salpêtrière, Paris, France

SUMMARY Three patients showed dramatic psychic akinesia after recovery from toxic encephalopathy. They had no or only mild motor disorders. The spontaneous psychic akinesia was reversible when the patient was stimulated, as if there was a loss of self psychic activation. Intellectual capacities were normal. Two patients had stereotyped behaviours resembling compulsions. In all patients CT cans showed bilateral lesions in the basal ganglia, mainly within the globus pallidus.

Mental disorders occur very frequently in basal ganglia diseases, such as Wilson's, Parkinson's, Hallervorden-Spatz diseases, Huntington chorea, and progressive supranuclear palsy... The clinical picture of these mental disorders however, are extremely varied; they consist of psychiatric disorders, personality changes and often various degrees of intellectual deterioration, but this last feature can be absent. In spite of this diversity a peculiar clinical picture characterisation by slowing down of thought processes, inertia and memory disorders has been identified in several degenerative diseases, and has been described under the term of "subcortical dementia".1,2 Degenerative lesions are generally not limited to the basal ganglia, so eventual role in mental processes cannot be established. Contrarily, in the motor control field, there is much anatomical, physiological and clinical evidence (the two latter largely from the study of Parkinson's disease), leading to the conclusion that basal ganglia should be regarded as organs of motor control, as recently emphasised by Marsden.3

We have recently drawn attention to a psychic picture,4,5 possibly the consequence of bilateral subcortical lesions in the basal ganglia, which was characterised by pure psychic akinesia without permanent dementia, and by pseudo-obssessional activities. We now report three similiar cases, which, in combination with some clinical and anatomical data in the literature, may furnish some support to the hypothesis of a role played by the basal ganglia in mental processes.

Case reports

Case 1: In 1968, Mr V, a 41-year-old healthy man was stung on the left arm by a wasp. He immediately sustained a convulsive coma for 24 hours, then adopted intensive choreic movements (which were alleviated by thio-properazine), and impairment of gait. These extrapyramidal symptoms diminished over several months. Then, and during the twelve following years he appeared to be a mild dement, He was evaluated by us in 1980, and was at this time not receiving drugs. All his activities were dramatically reduced. He spent many days doing nothing, without initiative or motivation, but without getting bored. The patient described this state as "a blank in my mind". His affect was disturbed. When talking about family problems, sad or pleasant, he had an appropriate behaviour and gave signs of normal interest, but this attitude did not last and he became rapidly indifferent again. His fantasy life was poor, but dreaming was preserved. But when stimulated by external events, or more specially by another person he could perform quite correct complex tasks (for example, playing bridge). This fact was dramatically demonstrated by neuropsychological tests which showed intellectual capacities within the normal range (table).

Two years after the encephalopathy, he began to show stereotyped activities. The most frequent consisted in mental counting, for example, up to twelve or a multiple of twelve, but sometimes it was a more complex calculation. Such mental activities sometimes were accompanied by gestures, such as a finger pacing of the counts. To switch on and off a light for one hour or more was another of his most common compulsions. When asked about this behaviour he answered that he had to count... that he could not stop it... as that it was stronger than him... Once he was found on his knees pushing a stone with the hands; he gave the explanation that he must push the...
stone, and he used the hands because he experienced some difficulties in skilled movements with his legs. There was however no anxiety, and in his past history there was no suggestion of an obsessional neurosis. Personality evaluation was normal (Table).

Neurological examination showed abnormal movements. At the time of examination in 1980 choreic movements were very mild but voluntary movements were often brisk. He had a permanent facial rictus with some facial or mandibular movements somewhat resembling tics. With his finger movements it was difficult to distinguish between involuntary or “voluntary” activity associated with mental counting. Walking was a mixture of Parkinsonism and choreic disturbances.

Standard and sleeping EEG were normal, CT scans were performed in the orbito-metatal plane. Slices were taken every 3 millimeters in the basal ganglia area. An evaluation of lesion sites was made as precisely as possible after CT scan film magnification to real dimensions, by comparison with an atlas. The main lesions consisted of low density areas situated bilaterally in the internal part of the lentiform nucleus (fig 1). The rostral part of the nucleus seemed to be more affected than its caudal part. Some other small low density areas could be seen within the rostral right putamen and the head of the right caudate nucleus. No enhancement was noticed after contrast infusion. Mild ventricular enlargement was present.

Several drugs were systematically tried. Dopaminergic agents (agonist and antagonist), serotoninergic, cholinergic, noradrenergic and benzodiazepines were used. Most of this drugs had no or mild effects on the patient’s symptomatology. Then, clomipramine was given up to 250 mg/day (under cardiovascular supervision) and this drug induced a dramatic improvement. For the first time for twelve years the patient was able to take the initiative to drive a car, and to initiate talking. Speech fluency reduction and stereotyped activities, however, remained.

The patient died suddenly from massive inhalation of food. Necropsy was not performed.

Case 2: Mr D was 23-years-old when he sustained in November 1979 carbon monoxide poisoning confirmed by blood level measurement (3-2 mm%). He was examined for the first time by us in January 1980. Neurological examination was normal except for intellectual perfor-

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**Table**

<table>
<thead>
<tr>
<th>Patient 1</th>
<th>Patient 2</th>
<th>Patient 3</th>
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<tbody>
<tr>
<td><strong>Attention</strong></td>
<td><strong>Orientation</strong></td>
<td><strong>Orientation</strong></td>
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<tr>
<td>—correct orientation</td>
<td>—correct mental control (Wechsler memory scale sub-tests: 9/9)</td>
<td>—orientation to time and space correct</td>
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<tr>
<td>—correct mental control (Wechsler memory scale sub-tests: 9/9)</td>
<td>—forward digit span limited to 4 items.</td>
<td>—mental control (Wechsler memory sub-test: 4/9)</td>
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<td>—forward digit span: five items.</td>
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**Specific performances**

| Poor spontaneous speech; reduced fluency; correct naming; poor evocation of animal names (12 out of 60 seconds); story of Little Red Riding Hood incomplete; correct reading and spelling; handwriting disorder; I.Q.: 103 on the vocabulary test of Binois-Pichot (6). Correct calculation. No apraxia. Visuo-constructive activities: correct explorations; some blanks in the copy of Rey’s complex figure. | Verbal fluency reduced, correct naming, evocation of animal names correct; correct reading and spelling; correct handwriting. I.Q.: 116 on the vocabulary test of Binois-Pichot. Correct calculation. No apraxia. Visuo-constructive activities: good copy of Rey’s complex figure. | Language: the voice is low, fluency is reduced and talking is poorly informative. There is a lack of words, handwriting is incorrect. The patient could not perform the vocabulary test of Binois-Pichot. It must be kept in mind that the patient is of Russian origin. Calculation incorrect. No apraxia. Visuo-constructive activities: correct exploration; good reproduction of Rey’s complex figure. |

**Intellectual performances**


**Memory**

| Retention and learning impaired (Wechsler memory scale: 88) | Retention and learning impaired (Wechsler memory scale: 70) | Insufficient results in retention and learning (Wechsler memory scale: 97) but correct evocation of recent events and remote memories. |

**Personality evaluation**

| Rorschach test: the test does not show any indication of an obsessional strucrutation; low self control; there is however some indication of an organic psychosyndrome. Minnesota Multiphasic Personality Inventory (MMPI) —normal profile; mild signs of hysterical nature; depressive and anxious elements are absent. The personality is in the normal range, albeit poor. | Rorschach test: paucity of protocol, numerous banalities and stereotypes. MMPI: normal pattern. | Rorschach—answers are qualitatively good and personality seems to be well controlled. There are probably some conflicts which are underlined by difficulties in sexual identification. There is no depression. MMPI: non valid. |
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Fig 1 Case 1: CT scan, 42 mm above the orbito-meatal plane and schematic representation.

mance; memory and verbal fluency seemed deeply disturbed. The patient was examined again one year later. He had still severe disorders of memory and verbal fluency. However, intellectual performances were dramatically improved. He could perform complex tasks correctly and solve problems. But these faculties were largely underused. In the absence of external stimulation he lay for hours, eyes open, doing nothing. The contrast between his intellectual capacities and this inactivity was obvious in all aspects of his life. He talked only if asked, he took part in sport (he was a sports coach) only if stimulated by his wife, he went to visit friends only if invited by a phone call, and so on. His only spontaneous activity was of a routine nature like going out and getting the newspaper. His affect was impaired in the same way. If asked about the recent death of someone he cared about he cried sincerely, but if asked about recent events of his life he forgot the death and talked only about some political news. Stereotyped activities were not reported spontaneously by the patient, nor by his wife. But when questioned on this point, he admitted counting when he was alone with nothing to do; he counted from 1 to 20 again. To stop it he had to go out, or watch TV. This purely mental activity did not give him anxiety nor did its withdrawal. Besides this activity, there was no sign of an obsessionnal neurosis. Neurological examination was normal. Neuropsychological evaluation showed no deterioration except for a memory disorder (table).

EEG was normal. CT scan (orbito-meatal plane, slice thickness 10 millimeters) showed two low density areas almost symetrically placed in the internal part of the lentiform nucleus (fig 2). These low density areas were visible on one slice only. There were no other abnormalities.

Case 3: Mr P born in Russia in 1911 and living in France since 1933, suffered in March 1970 carbon monoxide poisoning. There was a short coma followed by several days of headache and confusion. This man, whose profession was that of an artistic painter but who also did the job of messenger, was unable to re-start his work. He attended for neurologic consultation in April 1970, after being fired from his job because he was too slow. He had a "marche à petits pas", a resting tremor, an extrapyramidal rigidity predominant on the left side. Akinesia was obvious and verbal fluency was markedly reduced. Intellectual processes were slow and the whole picture tended to give the impression of mental deterioration. He was, however, able to perform complex tasks on request. He was institutionalised and his status improved during the first year, both in the extrapyramidal syndrome as well as in intellectual performances. At first unable on admission to conceive and execute basic drawings, one year later he could produce elaborate and artistic drawings and paintings. The most striking feature lay in his dramatic passivity, his lack of initiative despite the fact that his motor and mental capacities were largely preserved. He stayed in a ward, spending most of the time doing nothing and he never attempted to leave hospital. If questioned on this point, he answered that he didn't know, or that an authorisation to leave was required, that he had no family... anyway, he didn't feel bored. His lack of initiative was, however, not total since he had some spontaneous activities, such as helping the other patients in eating and shaving, and sometimes he watched TV, or read the newspaper. He went out to walk in the park only if he had been actively encouraged. He was able to perform artistic paintings; but for years he painted the same landscape of moors and fens, and this several dozens of times. His affect was poor in relation to his solitude—when questioned about his biography, he evoked spontaneously with sadness the death of his mother or brother.
Neurological examination was normal. A mild motor akinesia was noticed. Neuropsychological evaluation is shown in the table. Language disorders were present, some of them due to the Russian origin of the patient.

The EEG was within normal limits. CT scan (orbito-meatal plane, slice thickness 10 mm) showed two bilateral abnormal areas with spontaneous high density, which corresponded to calcified lesions (fig 3) in the internal part of the lentiform nucleus. There was no other calcification. There was some degree of cortical sulci widening and of ventricular enlargement. Contrast medium infusion showed no enhancement. Laboratory data, including serum calcium phosphorus and parathyroid hormone levels were within the normal range.

Fig 2 Case 2: CT scan 40 mm above the orbito-meatal plane and schematic representation.

Fig 3 Case 3: CT scan 45 mm above the orbito-meatal plane and schematic representation.
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Discussion

PSYCHIC AKINESIA

The main disorder in our three patients was a spontaneous psychic akinesia, which led to a severe reduction of both mental and behavioural activities. Unlike patients with diseases which induce psychic and "motor" akinesia such as Parkinson disease, our patients had no reduction of their motor ability. Their normal or largely preserved intellectual capacities stood in contrast to their spontaneous psychic inertia. This was especially evident during neuropsychologic testing. The other symptoms of our patients were a defect in speech fluency, memory disorder (case 2 and 3), a decrease in affect and stereotyped activities (case 1 and 3). The psychic akinesia was less severe in case 3.

As shown by neuropsychologic assessment, there was no aphasia, apraxia, nor agnosia. Somewhat similar pictures have been described as subcortical dementia in neurological diseases such as progressive supranuclear palsy, Parkinson disease, Huntington's chorea. This terminology does not imply, however, that any cortical involvement is absent, but that lesions presumably predominate at subcortical and basal ganglia level. Indeed the psychic disorder often encountered in progressive supranuclear palsy is similar to that of our patients. According to Albert et al's description, it includes: (1) memory disorder, (2) slowing of thought processes, (3) personality changes with inertia and apathy, (4) impaired ability to manipulate acquired knowledge. The neuropathological changes of progressive supranuclear palsy affect primarily the upper brain stem, but they frequently also involve the basal ganglia, and especially the pallidum.

In Parkinson's disease the mental disorders have been widely investigated. They consist mainly of depression, a variable degree of impairment of intellectual function, personality changes, with sometimes a slowing down of mental process. This slowing down of thoughts has been described in the literature under various names, such as bradyphrenia, mental viscosity, or psychic akinesia. This state was described as follows by Mettler: "with hypokinesia and rigidity appear a slowed reaction time, a loss of mental speed and agility, a certain deliberateness and indecision or suspension of decision, without any real evidence of impairment of intellectual capacity". However reversibility of psychic akinesia by external stimulation was not noticed. Though some demented Parkinsonian subjects may have cortical lesions, it has been shown that the slowing down of mental processes was best correlated with subcortical dysfunction. Similar psychic akinesia was also described in post-encephalitic Parkinsonism, where cortical involvement is usually small. But in Parkinsonism, as in progressive supranuclear palsy there is an obvious reduction of motor abilities, in contrast with our patients who had no or only mild extrapyramidal symptoms and thus displayed a pure psychic akinesia. Such descriptions of pure psychic akinesia, reversible under external stimulation, are rare in the literature. They are however mentioned as pure mental sequelae of encephalitis lethargica (Bleuler personal communication, and Naville). This latter author gave the following description: "these patients if they are not stimulated at every moment stay doing nothing as if they have lost their psychic tone. . . . This syndrome may be reversible by an external stimulus as it was able to compensate for the lack of internal stimuli". Unfortunately no pathological data are available in such observations. It can be noticed however that in sequelae of encephalitis lethargica, the usual lesions are primarily located in brain stem, basal ganglia including pallidum, the cortical changes being generally moderated or absent.

From another point of view there are some similar features in the clinical picture of our patients and symptoms of an impairment of frontal lobe activity. Patients with frontal lobe lesions also display a slowing down of thoughts, a decrease in behavioural activities, apathy, indifference. These similarities between subcortical dementia and frontal lobe syndrome were pointed out by Albert. This comparison is also supported by experimental studies in the monkey which showed that lesions of the head of the caudate nucleus can induce behavioural disturbances very similar to those induced by a prefrontal lesion. As far as our cases are concerned there are however strong differences, since when stimulated our patients were able to have and to maintain normal intellectual activities or behavioural programmes, whereas the maintenance of attention upon a task is very difficult in patients with frontal lobe syndromes.

This reversibility of the psychic akinesia under stimulation was so dramatic in our patients that it is not possible to apply the term of dementia to them. Besides mnesic impairment (cases 2 and 3) they had no permanent disorder of specific intellectual function. It appears as a loss of psychic self-activation concerning all mental activities, cognitive and affective including fantasmatic life. This defect in spontaneous internal activation could be compensated for by external stimulation. Some psychiatric conditions also present a more or less reversible psychic akinesia, such as hebephrenia or some depressive states.
COMPULSIVE-LIKE ACTIVITY

The existence of stereotyped behaviour in patients 1 and 2 represents another dramatic aspect of these cases. They consisted mainly of a counting activity, purely mental or paced by finger movement. In patient 1 they could occupy all the time he spent alone, unstimulated. These activities may be interpreted as compensating activities which invade an empty, aimless mind, deprived of more elaborate purposes. Such an interpretation is in agreement with the dynamic theory of Janet who described repetitive activities as the lower level of psychic life, due to a decrease in "tension psychologique".

In several aspects this repetitive behaviour strongly resembled compulsive acts. Patient 1, when questioned on his activities answered that he had to do so; when counting he had to reach some magic numbers or multiples of these numbers; the second patient made attempts to withdraw himself from these behaviours by focusing his attention upon TV or a newspaper. However in spite of the compulsive appearance of these activities and some struggle against them, patients did not feel any anxiety when they tried to withdraw. In their daily life they did not show any anxious thought, or permanent doubt such as that showed by obsessional subjects. Nothing either in their histories or in the recent assessment of their personality suggested an obsessional neurosis.

It is of interest that in some neurological diseases obsessive—compulsive behaviour has been reported and that they were thought possibly to be due to basal ganglia impairment. In numerous cases of Parkinsonism, most of them being postencephalitic cases, obsessive—compulsive activities were described such as arithmomania, vocalisations, or purely obsessional thoughts. These behaviours often occurred in short crises lasting some minutes and could be accompanied by motor disorders such as oculocorporeo-lingual fits and by personality changes such as emotion—aggressivity but mainly anxiety. Some cases, however, are very similar to our patients, since obsession—compulsions are described as occurring without modifications of the emotional state, as a substitution phenomenon triggered by an inner blocking of thoughts. The association of obsession—compulsions and tics can be present in Gilles de la Tourette syndrome. An organic background at the basal ganglia level has been suspected in this disease on the basis of anatomical, clinical and pharmacological findings.

THE ROLE OF PALLIDAL LESION

On CT scan of the three patients lesions were present in the deep forebrain at the basal ganglia level. In the following discussion, we do not imply that our patients had no other brain lesions, such as in the cerebral cortex, but we think that there is some reason to suggest that these basal ganglia lesions play the most important role in the mental symptomatology: (1) the lesions are presumed to be located in the pallidal area and the patients suffered from diseases known to determine lesions in that area. (2) several data in literature suggest or demonstrate that pallidal lesions may be followed by mental changes resembling to psychic akinesia.

CT scan analysis by magnification and comparison with an atlas suggest that the bilateral lucencies were located in the internal part of the lentiform nucleus, that is the pallidal area. This precise site of lesion is frequently observed on neuropathological examination of brain from subjects who died from carbon monoxide intoxication. It is thus widely assumed that the bilateral lucencies discovered on CT scan in the same aetiology are actually located in the globus pallidus. Lesions in case 1 seem to be more lateral, presumably overlapping the putamen. Necrotic lesions with this localisation have been reported in wasp sting encephalopathy from pathological material. In both diseases, however, the white matter and/or cerebral cortex can be involved but, if it was present in our patients, such damage was too subtle to be seen on CT scan.

As previously outlined several diseases which include a pallidal impairment can give a psychic akinesia, that is progressive supranuclear palsy, Parkinson's disease sequelae of encephalitis lethargica. More interestingly bilateral subcortical lesions performed for the management of some Parkinsonian symptoms have induced psychological changes somewhat similar to our patients' disturbances. It is sometimes difficult to get a clear picture of the effects of bilateral operations, as several reports do not distinguish between pallidal and thalamic lesions on one hand and between unilateral and bilateral lesions on the other hand. It appears however that the mental changes are more pronounced when the pallidum is involved. Unilateral pallidal lesion may induce transient changes, but bilateral pallidal lesions, or pallidal and controlateral thalamic lesions, can induce more durable changes. These changes are described as inertia, lack of drive, decrease of emotional—motivational behaviour or as a psycho-organic syndrome with sometimes compulsory activities. They are summarised by Ricklan and Levita as a non specific decline in the maintenance of energy level and initiative which contrasts with the lack of changes on specific tests. Speech impairment is often reported. These symptoms seem to be less severe than in our patients, but stereotaxic lesions are usually small, affecting only a part of the globus pallidus. On the contrary, however, a case of
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almost total reduction of psychic and motor activities is reported after such bilateral surgery with post mortem anatomical study which showed large bilateral pallidal lesions.40

Further support for a role of pallidum in psychic processes can be obtained from the study of its connections. Several connections have been established between the globus pallidus and some brain areas which are not acknowledged as motor structures. On the input side, it has been demonstrated in the monkey41-42 that the rostral part of globus pallidus receives afferents from rostral and medial parts of neostriatum. The rostral neostriatum receives predominantly, though not exclusively, fibres from prefrontal cortex.43-45 This was also suggested by behavioural studies from Rosvold.18 It is thus possible that in the monkey and in man some part of the globus pallidus and rostral neostriatum functions primarily with the prefrontal cortex, as shown in the rat.46 This fact can account for an anatomical basis for similarities between some aspect of subcortical dementia and frontal syndromes. Moreover, in the rat, a strong limbic input (from amygdala and hippocampus) has been demonstrated on the rostral neostriatum.47-48

The efferent pallidal fibres which leave the basal ganglia arise from the medial segment of globus pallidus. A large “motor” outflow is directed toward the ventral lateral–ventral anterior thalamic complex. But this medial segment of GP sends also efferents toward intralaminar thalamic nuclei and then to diffuse areas of neocortex, toward the brain stem and toward the habenular nuclei.49-52

It has been suggested from investigations in the rat52 that fibres destined for the habenula arise primarily from the rostral half of the entopeduncular nucleus—the rat’s equivalent of medial globus pallidus and that this part of the nucleus might be termed the “limbic pallidum”. The pallidal organisation in its output elements seems more complex in primates.54-55 A central zone can be described where most neurons send axonal branches to both the thalamus and the mid-brain, and a peripheral limbic zone which encroaches upon lateral hypothalamus and whose cells project only to the habenula. These projections may be of great significance since they mean that the striatal efferents mediated by the medial globus pallidus have limbic interrelations. If these habenular connections are dense in animals, their importance remains however to be determined in man.

Besides the heterogeneity of its connectivity the globus pallidus proper is crossed or bordered by several systems of fibres in passage, and is embedded in a peripallidal area which contains functionally different cells. The extension of the lesions to the peripallidal area cannot be accurately established from CT scans in our patients, but such a possibility has to be taken in account. Thus several other hypothesis may be formulated, as contributing to the generation of the mental symptoms: (1) slight encroachment upon the anterior limb of the internal capsule (2) involvement of the ventral extension of the pallidum within the substantia innominata, the functions of this recently described ventral pallidum remaining largely unsettled.46 (3) Interruption of ascending fibre bundles especially the catecholaminergic pathways which run in the vicinity of globus pallidus57 (4) lesion of the cholinergic cell clusters, such as the basal nucleus of Meynert which lie just beneath the globus pallidus.58-59 These cells are responsible for the extrinsic cholinergic innervation of the neocortex and their degeneration is currently the subject of a great research interest since it occurs in Alzheimer dementia (see for review ref 60). However the clinical consequences of impairment of these cells remain unknown.

Such a complexity of the pallidal area helps to explain the complexity of the clinical manifestations which can be observed in the case of lesions in this site (see ref 61 for detailed review). It is somewhat surprising that damage to this area can induce diversified motor disorders such as rigidity, dystonia,50-52 postural disturbance53 or pure Parkinsonism.27-24 The simultaneous occurrence of motor and psychic impairment is not expressly mentioned in the literature concerned with the consequences of pallidal damage.61 It seems conceivable, however, that psychic akinesia may be present in some cases, but may be very difficult to discern when speech and gesture are severely impaired. Finally pallidal damage can exist without neuropsychological symptoms.61 The possibility of pure motor disorders and conversely of pure psychic disorders as in the present cases, suggests that the lesions may not involve equally the different anatomical systems located in the pallidal area. Furthermore a disorder in this area should be considered in research on several psychiatric conditions, such as hebephrenia, obsessional neurosis or severe depression.

References


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