Arch. Psychiat. Nervenkr. 220, 1—7 (1975)

Akinetic Mutism Due to Glioma of the Midline

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Received April 9, 1974

Summary. The case described presents all clinical features for the definition of the true "akinetic mutism" syndrome. The anatomical examination revealed a polymorphous glioblastoma involving the structure of the midline, and particularly the rostral region of the gyrus cinguli, corpus callosum and the septal areas.

Key words: Akinetic Mutism by Midline Glioblastoma — Clinic and Pathology.

Zusammenfassung. Der beschriebene Fall weist alle klinischen Merkmale auf, die den echten „akinetischen Mutismus" charakterisieren. Die anatomische Untersuchung zeigt ein polymorphzelliges Glioblastom der vorderen Mittellinie, vor allem im rostralen Gebiet des Gyrus cinguli, Balken und Septum.

Schlüsselwörter: Akinetischer Mutismus nach Mittellinienglíom — Klinik und Pathologie.

The term "akinetic mutism" is often given an incorrect or too extensive meaning. It may, therefore, be of interest to recall the original definition of Cairns *et al.* (1941) (an observation concerning a case of epidermoid cyst of the Rathke's pouch).

"The patient sleeps more than normally, but he is easily roused. In the fully developed state he makes no sound and lies inert, except that his eyes regard the observer steadily, or follow the movement of objects, and they may be diverted by sound. Despite the steady gaze, which seems to give promise of speech, the patient is quite mute, or he answers only in whispered monosyllables. Oft-repeated commands may be carried out in a feeble, slow and incomplete manner, but usually there are no movements, struggling, or evidence of negativism. Emotional movements also are almost in abeyance. A painful stimulus produces reflex withdrawal of the limb and, if the stimulus is sustained, slow feeble voluntary movements of the limb may occur in the attempt to remove the source of stimulation, but usually without tears, noise, or other manifestations of pain or displeasure.

The patient swallows readily but has to be fed. Food seen may be recognized as such, but there is evidently little appreciation of its taste and other characteristics; objects normally chewed or sucked may be swallowed whole. There is total incontinence of urine and feces."

Case Report

The present observation concerns a woman (Frieda J. . .), aged 62 years, who was admitted to the Psychiatric Clinic of the University of Berne, suffering from incoercible vomiting, without nausea, lasting several weeks.

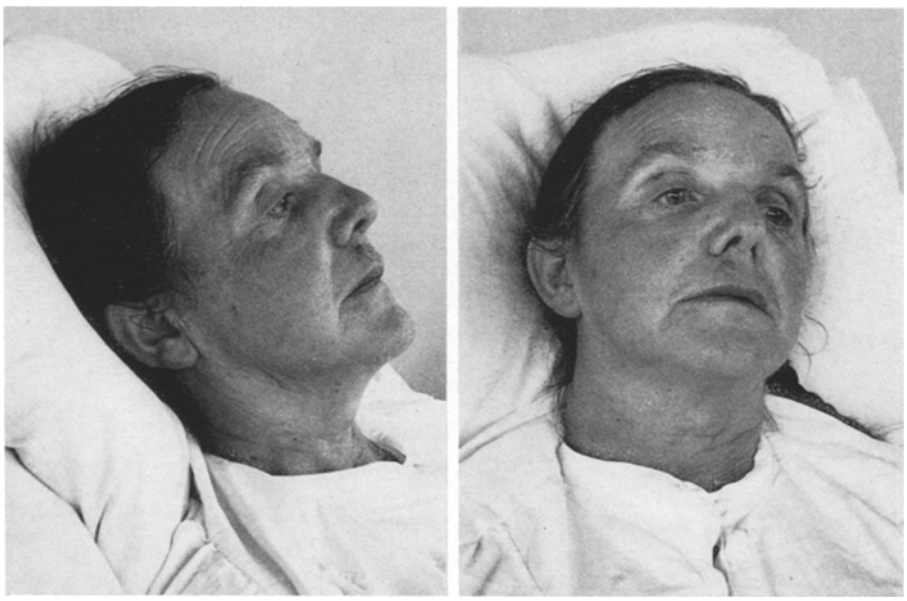

Fig. 1. The case described at the stage of total psychomotor inertia (akinetic mutism)

Three months previously the patient had begun to complain of violent fits of coughing, and sphincter dysfunctions. During the same period recent memory disorders appeared.

One week prior to admittance vomiting had become so frequent as to render adequate feeding impossible. At the same time marked general apathy appeared accompanied by increasing indifference to domestic surroundings.

When admitted to the Clinic a marked mental deterioration was noted, accompanied by indifference to the environment, emotional withdrawal and fixation amnesia with a tendency to confabulation.

The general examination was negative, except for a small perimalleolar oedema.

The neurological examination revealed a hoarseness of the voice, no dysarthria, isocoric central eucyclic pupils with a torpid reaction to light, no defect in the extrinsic ocular motility, a positive bilateral grasping reflex, a little adiadochokinesia in the right limbs, and a Babinski sign on the left.

There were no serious defects in the motor and sensory functions in the limbs, nor any significant changes in proprioceptive reflexes.

During the first month of hospitalization the psychomotor inertia increased, the bilateral grasping reflex became more pronounced, and oppositional resistance appeared in the four limbs. A suction reflex also became apparent.

The patient remained immobile in bed and presented a state of total inertia except for the eyes which from time to time followed movements in the environment, thereby showing that she was alert (Fig. 1). Only repeated verbal stimuli elicited a reply in the form of incomprehensible monosyllables.

E.E.G. The recording presented slow diffuse bilateral changes with focal signs in the left frontal region consisting of slow sharp waves of great amplitude spreading to the opposite side.

Angiography of left common carotid artery: Posterior cerebral artery not visualized. The anterior cerebral artery and its pericallosal branch were on the midline, but the latter followed a winding course.

The calloso-marginal artery was noticeably stretched, deviated upwards, and at a greates distance than usual from the pericallosal artery.

In a late arteriographic phase a network of abnormal vessels, arousing from a roundish mass in the region of the gyrus cinguli and the corpus callosum, was observed. The abnormal vessels were irregular in diameter and appeared to be situated in the peripheral zone of the mass. At the lower edge of it the abnormal vascularization had a nodular appearance, while at the upper edge it was arranged radially.

Drainage took place through small arteriovenous fistulae that ran either towards the deep veins and inferior sagittal sinus or towards the veins of the hemispheric convexity.

Angiographic diagnosis: infiltrating fronto-medial neoformation invading the corpus callosum, mainly on the left side.

The patient was submitted to roentgen therapy.

Two months after admission the patient presented difficulty in deglutition and developed a left central facial palsy.

Disturbances of the blood pressure regulation, cardiac arrythmia and generalized convulsions were also noted.

Food was administered through an oesophageal tube.

A downhill course followed. However, in spite of the total absence of any initiative, either motor or verbal, the patient continued to give the impression of following with an alert look every activity taking place around her.

After eight months of clinical course the patient developed a fatal bronchopneumonic infection.

Macroscopic Examination of the Brain

Total weight 1200 g.

The cortical convolutions were distinctly flattened and the sulci collapsed. A tentorial impression was clearly visible in the hippocampal gyrus on both sides.

The brain was fixed "in toto" in 10% formalin and coronally sectioned.

In a section 5 cm caudal to the frontal poles (Fig. 2) a massive *bilateral* infiltration had spread into the *corpus callosum* and the *gyrus cinguli* and also extended into the depth of the frontal lobes.

The neoformed greyish-yellow web was relatively compact and slightly vascularized. Due to the presence of peripheral necrotizing areas, its limits were not clearly defined.—In a more caudal section, at the level of the anterior portion of the corpus *striatum* (Fig. 3), the *septum pellucidum*, the *corpus callosum* and the adjacent pericallosal regions were completely invaded.

The *lateral ventricles*, and in particular, the left one, were almost completely filled up. The median structures appeared to be displaced less than one centimetre to the right.

In a frontal section at the level of the *nucleus amygdalae* (Fig. 4), the neoplastic infiltration was no longer visible macroscopically. In this and the subsequent caudal sections the only macroscopic changes were a venous congestion and a conspicuous tissue oedema.

Microscopic examination of the brain revealed a *polymorphous Glioblastoma*, which showed necrotic masses in the centre.

Discussion

This was a case of glioblastoma that had invaded the septum pellucidum, the corpus callosum and the cingular gyri and also penetrated partially into the depths of the frontal lobes.

The main clinical features corresponded to a typical "akinetic mutism" syndrome.

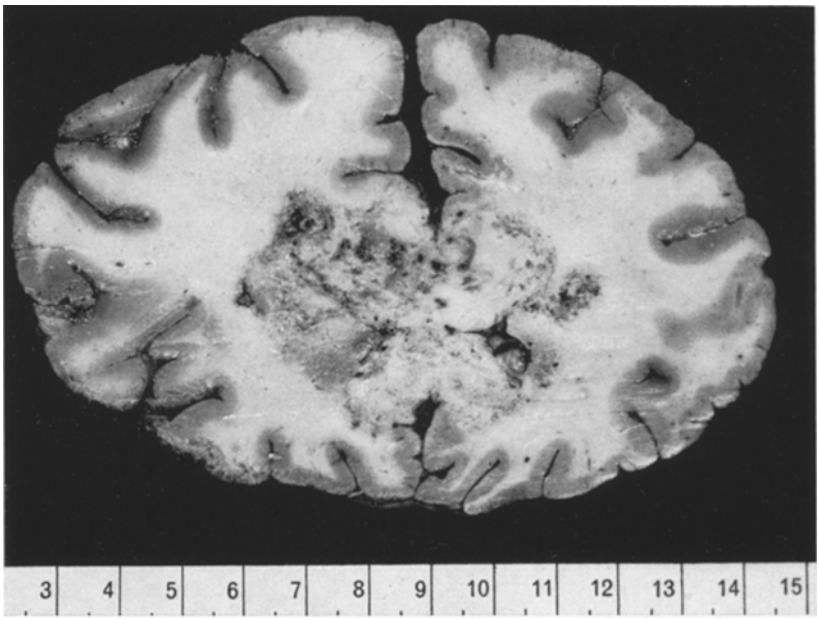

Fig. 2. Coronal section of the brain at the level of the genu corporis callosi. Massive gliomatous infiltration into the genu and gyrus cinguli

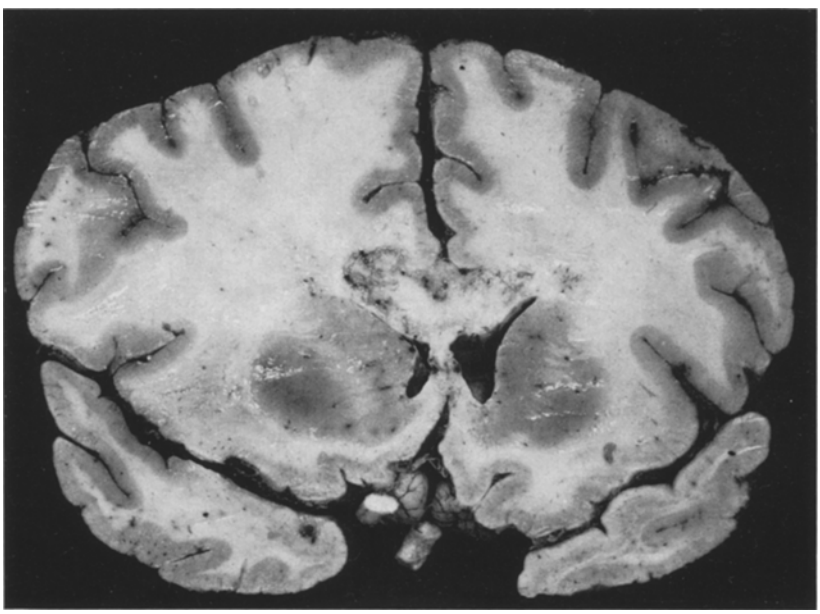

Fig. 3. Coronal section of the brain at the level of the rostral portion of the neostriatum. Gliomatous infiltration of the corpus callosum and septum pellucidum

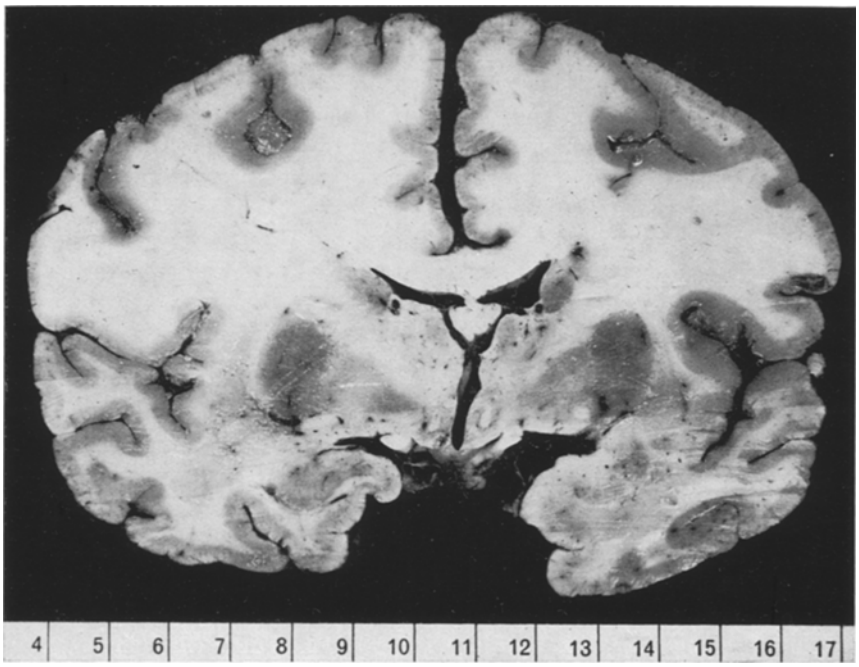

Fig. 4. Coronal section of the brain at the level of the amygdala. No traces of tumor; cerebral oedema

The original description of Cairns *et al.* was followed by a very large number of observations of "akinetic mutism". In most cases, however, the lesions were too extensive or multiple, therefore preventing a well-defined anatomo-clinical relationship. We would draw attention to the diffuse lesions following carbon monoxide poisoning (Cravioto *et al.*, 1969) and the aspecific cerebral lesions caused by general organic decline (Segarra and Angelo, 1970).

As previously explained the term "akinetic mutism" has recently been used too extensively and sometimes improperly. This applies particularly to the conditions described by Segarra and Angelo as "pseudo-akinetic mutism" (mutism of an extrapyramidal origin, "sentient mummy syndrome", the "apallic syndrome").

Various *diseases of the basal ganglia* may terminate in an extreme condition of akinesia, rigidity and mutism. They include necrosis of the putamen and caudatum described by Marinesco (in Segarra and Angelo), cases of pallidal necrosis (Denny-Brown, 1962), the purely pallidal lesions in carbon monoxide poisoning and some stereotaxic pallidal lesions in Parkinson's disease.

The "*sentient mummy syndrome*" ("locked-in syndrome" of Plum and Posner, 1966) is caused by multiple softening in the pons, affecting the cortical descending pathways to the brain stem nuclei and spinal cord. Ocular movement is preserved and can be used by the patient as a means of communication.

None of the observations of Cravioto *et al.* conform to this type, nor does the first case described by Fang and Palmer (1956). It is obvious that the use of the term "akinetic mutism" for a condition which is a purely motor defect will lead to

a confusion in terminology. Kemper and Romanul (1967) stress the necessity of separating the two conditions.

The "*apallic syndrome*" is generally associated with extensive cortical damage. The most frequent causes are anoxia, carbon monoxide poisoning, closed head injuries, and diffuse encephalopathies. Distinction from the true "akinetic mutism" is facilitated by the existence of serious changes in muscle tone and posture. The observations described by Jellinger *et al.* (1963) are of this type.

In the true "*akinetic mutism*", akinesia and mutism appear without paralysis or any serious sensory-motor disturbance, and without diffuse damage of the cerebral cortex and basal ganglia.

Causal lesions of the true "akinetic mutism" have two possible locations, either the region of the median mesodiencephalic junction or the cingular gyri.

1. Akinetic Mutism: Meso-Diencephalic Median Lesions. In the most typical cases, following the occlusion of the retromammillary branches of the posterior cerebral artery, the ischemic necrosis inevitably affects the periaqueductal grey matter and the median reticular substance at the level of the meso-diencephalic junction.

Rostral extension of the lesion into the medial thalamic nuclei, subthalamus and hypothalamus, as well as its caudal extension into the midbrain, may vary from case to case (Angelergues *et al.*, 1957; Barraquer-Bordas *et al.*, 1967; Brage *et al.*, 1961; Castaigne *et al.*, 1962; Façon *et al.*, 1958; French, 1952; Haase and Luhan, 1952; Kroh, 1964; Lechi and Macchi, 1967, 1974; Lhermitte *et al.*, 1963; Riser *et al.*, 1957; Segarra and Angelo).

The typical disturbance of the state of consciousness (with which hypersomnia is frequently associated) coexists with variable neurological symptoms arising from thalamic, subthalamic, hypothalamic and mesencephalic damage.

2. Akinetic Mutism: Cingular Lesions. In cerebro-vascular pathology this condition follows an ischemic bilateral lesion in the regions supplied by the anterior cerebral artery. Involvement of the anterior cingulate regions (area 24) appears to be particularly significant.

Observations of this type have been reported by Faris (1967), Nielsen and Jacobs (1951), Amyes and Nielsen (1955), Barris and Schuman (1953), Manghi *et al.* (1959).

In these cases the disturbance of the state of consciousness may be even less characteristic (i.e. frontal apathy, confusional or confuso-demential states, etc.). However, the preserved alertness observed in them contrasts with the torpid condition of the "akinetic mutism" of a meso-diencephalic origin.

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