AFFECTIVE EPILEPSY*

BY

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In the early part of this century several writers (Oppenheim, 1906 ; Bratz, 1907 ; Dana, 1907, and others) reported fits which were neither epileptic nor hysterical but which, they believed, were provoked directly by external stimuli, such as slight excitement or over-exertion causing an emotional change. The term "affective epilepsy" was introduced by Bratz and Falkenburg in 1904 to describe these fits, which often resembled the prodromata and aura of true epilepsy but were accompanied by outbursts of emotion and not by epileptic convulsions. When it was found that such cases often turned out to be of organic origin interest in them gradually declined. Now that the psychological aspects of epilepsy are again receiving much attention from clinicians and investigators, however, it may be worth while to reconsider this syndrome of affective epilepsy and in particular to study a case in which it played a prominent part.

Case History

The patient was first admitted to the Valerius Clinic in 1941 at the age of 23. For two years he had suffered from minor fits, which had become more frequent shortly before his admission. There were no convulsions. During these short fits the eyes rolled, or he had a vacant look, or a transient moving of the lips. Consciousness was lowered during the fit, and there was aphasia for about two minutes following the attack. There was amnesia for the fits, although he would remain sitting or standing. There were no complaints of headache, vomiting or vertigo, nor of disturbance of micturition. There were no personality changes.

Neurological examination in November, 1941, showed impaired motor function of the right facial nerve and divergent strabismus to the left. Air-encephalography showed that the anterior horns and, even more distinctly, the diencephalon were displaced to the right. The left anterior horn was displaced slightly upwards, and was slightly flattened. The posterior horns were normal.

Clinical data and ventriculograms indicated that the cause of the displacement must lie basally on the left, rather far forward, almost at the level of the foramen

of Monro. There was very little functional impairment. A frontal trepanation was made but inspection of the orbital roof and olfactory nerve showed no trace of tumour. Puncture met with a deep resistance, situated not quite frontally. It was therefore not possible to carry out a radical frontal lobectomy, and there was no point in risking functional impairment for the purpose of exploration. Only subtemporal decompression was carried out, with post-operative x-ray treatment. The patient was discharged home, at first quite well ; he gradually became more and more emotional, tired easily, and was infantile in affect.

He was admitted again in November, 1946. For the past year his fits had been accompanied by a curious emotional discharge. After slight excitement, such as entering the lecture-room or sitting in the screen-room, there would be a sudden lapse of fixation in his gaze, a vacant look, a transient moving of the lips. This would be followed by a period of slightly lowered consciousness and then by a violent emotional discharge. He would suddenly start to deliver a speech in a pseudo-language, with mimicry and gestures to match. If the content and sound of his words were disregarded, the affective expression was within normal limits and the modulation of his speech was normal. He would utter incomprehensible words, gesticulating violently as if trying to persuade someone ; clench his fists and pound on the table as if furious. A torrent of words would be released, consisting of apparently meaningless syllables. This would last for about three minutes, towards the end of which he would speak in a lower and lower voice, looking to the left. Finally he would say a few syllables in a wheedling tone, smile contentedly, and the fit would then be over. He would immediately recognize his environment and speak sensibly to those around him. During the emotional outburst he showed a stereotyped automatism of behaviour, but appeared to be fully conscious and in full possession of his faculties, though not normally in touch with his environment.

Neurological examination revealed an end-position nystagmus on the right as well as on the left. Arm and leg reflexes were a little brisker on the right than on the left but there were no pathological reflexes. The diencephalon appeared normal, but both on the left and on the right there was a slightly choked disc. Another air-encephalogram revealed an obstruction of the left interventricular foramen.

From January, 1947, he had irradiation treatment,

* Based on a lecture delivered at the Maudsley Hospital, London, in March, 1952.

FIG. 1.—The distended anterior horn of the left ventricle is visible. The tumour is localized on the basal side, both right and left.

FIG. 2.—This section shows a diffuse mass of tumour in the median region, with a cyst in its centre.

FIG. 3.—The section shows the markedly thickened columns of fornices and bodies of fornices as far as the mammillary bodies.

with some initial success, and he was discharged in February, 1947.

On his third admission, in July, 1949, he presented a puerile euphoria and signs of slight deterioration. An E.E.G. showed slow waves, frontal and temporal, on both sides. The most striking feature of his clinical behaviour was that he still showed occasional very

violent emotional outbursts. The neurological picture was unchanged.

The patient died in October, 1949, with signs of increased intracranial pressure. There was a gradual lowering of consciousness and he was subcomatose for some days before he died. The diagnosis was the same as eight years earlier: fronto-temporal tumour confirmed by neurosurgical operation.

The post-mortem examination showed that the convolutions of both hemispheres were markedly flattened. Adhesions with the dura were found in the left temporo-parietal region at the site of the old exploratory operation. The extent and type of the tumour are shown by Figs. 1 to 8.

To sum up the histopathological findings, the process extended throughout the basal subcallosal regions of the frontal lobes, the septum pellucidum, the fornices and the hippocampus, extending to the hypothalamus, the thalamus, the lenticular nuclei, and the cortex of the insula. The cingulate gyrus showed diffuse gliosis not clearly continuous with the main tumour mass. The outstanding feature was that the proliferation was practically confined to the rhinencephalon whereas the neopallium had suffered little.

Discussion

The initial problem arises from the emotional outburst. Emotion is a dynamically interrelated whole, at once physiological, psychological, and social. It is in a normal person a highly integrated self-regulating structure, connected with continuous conscious experience. It is not possible to study the phenomena of emotion without regard to the well known cerebral systems upon which they depend. This patient's clinical behaviour could be attributed to a complex of reverberating circuits, occasionally exhibiting very violent emotional discharges in which he availed himself of the various possibilities of expression in speech, face, and gesture. In this we have to distinguish between the expression of emotion and the experience of emotion.

In 1937 Papez pointed out in his fundamental study, "A Proposed Mechanism of Emotion", that the hippocampal formation with fornix, hypothalamus, anterior thalamic nuclei, and cingulate gyrus, together with their connexions, constituted a self-regulating system, accurately dealing with feeling tone as well as participating in emotional expression. However, though the patient was under my supervision from 1941 till 1949, the application of this hypothesis to his disorder did not occur to me. I concluded on his first admission that the cause of the displacement evident in the ventriculogram must be a tumour seated basally on the left,

FIG. 4.—The whole hippocampus and the cortex of the hippocampal gyrus are affected. The lenticular nucleus situated above them is completely filled up by neoplastic proliferation; the cortex of the insula, the thalamus, the hypothalamus, and the fornix also show proliferation. The cingulate gyrus is clearly affected, whereas the corpus callosum is not.

and my diagnosis remained the same when he died eight years later.

There are many indications that the rhinencephalon is largely concerned with emotional functions. This phylogenetically old part of the brain appears to be so strategically situated as to be able to correlate every form of internal and external perception. And, in contrast to the neopallium, the rhinencephalon has many connexions with the hypothalamus; though our intellectual functions are carried on in the newest and most highly developed part of the brain, our affective behaviour continues to be dominated by a relatively crude and primitive system.

The medial wall of the cerebrum is physiologically integrated with the hypothalamus and the lateral wall with the dorsal thalamus. Moreover, obsessive oral utterances can arise from irritation of the cingulate gyrus. In the clinical picture presented by our patient, we are justified, therefore, in suspecting irritation and radiation as consequences of morbid processes in the hippocampus and cingulate gyrus. The question of what our patient was feeling is irrelevant: as Bard (1934) showed, only the cortex is essential to the subjective experience of emotion.

FIG. 5.—The hippocampus on the right side in the same cut surface has a slightly swollen appearance. The thalamus is possibly slightly enlarged.

FIG. 6.—Nissl staining reveals that the right hippocampal region contains an excessive proliferation of cells as well as the thalamus and the hypothalamus, while there is unmistakable evidence of this process also in the fornix.

FIG. 7.—Glioblastoma multiforme. There are occasional masses of vascular proliferations, sometimes showing a similarity to glomeruli—"Scherer coils".

Lesions in the hippocampal formation or, to put it otherwise, in the Papez circuit, may intensively affect the expression of emotional life (anxiety, apprehensiveness, rage, and terror). The question arises whether patterns of emotional discharge may lead to excessive expression if they are not controlled by the neocortex, and whether release of these discharges will be repeated whenever stimulation occurs in the rhinencephalon. Here autochthonous stimuli might have arisen from the pathological glia.

In this case there are arguments for the condition being a developmental disease, and for its being a new growth. It is difficult to determine the original site of the process. Some displacement existed in the anterior part of the left basal region, at the approximate level of the foramen of Monro. The puncture needle met resistance immediately posterior to the frontal region.

Similar tumours have been described. Ostertag (1951) classifies them under a separate heading, gliomata of the allocortex. They are presumed to develop on the basis of developmental disorders. This theory is substantiated by the simultaneous existence of persistent embryonic fissures in the surface of the thalamus and ependymal changes, associated with cortical changes in the form of myelinization gliosis, proliferation of satellite cells, and persistence of the embryonic granular layer.

The olfactory parts of the brain are always predominantly affected.

In our case the gliosis in the cingulate gyrus is suggestive of a developmental disorder. The adhesion of the frontal hemispheres on the basal side of the longitudinal fissure is also reminiscent of a malformation. But the intensive proliferation of glia conveys a strong impression of an independent blastomatous process. There is little to support the diagnosis of a diffuse periaxial encephalitis. The naked-eye picture of the hippocampus, which is twice its normal size, indicates a new growth, which is corroborated by the microscopical findings.

So I would prefer to ascribe these diffuse neoplasms to a faulty "ability" of the glial tissue, reminiscent of a systemic disease, such as reticulosis. This view corresponds to the one held by Schwartz and Klauer (1927) who, after reporting their case, concluded :

"There actually exist blastomatous proliferations of the glial apparatus, arising all at once throughout the system—or in its major parts—from the innumerable autochthonous elements."

In spite of the marked increase of pathological glia, the structure of the cerebral system remained intact in many places. Axis cylinders have rarely been impinged upon, myelin sheaths more, though to a lesser extent than in diffuse sclerosis. We are faced, then, with a systemic disease in the rhinencephalon. But on the other hand the proliferation of glia is, for instance in the left lenticular nucleus, very intensive and without any respect for the pre-existent structure ; there were patches of necrosis

FIG. 8.—Nissl's "astrocytome à grandes cellules".

and vascular proliferations. It might then be inferred that this process started as a fairly benign, slowly growing spongioblastoma and macrocellular astrocytoma, which finally developed into a glioblastoma.

Summary

Summarizing, we have a remarkable process extending through the entire rhinencephalon in a 31-year-old man, who had had fits for 10 years and, during the last four years, symptoms of affective epilepsy. The three-fold interest of the case lies in the clinical value of the syndrome as a guide to the pathological diagnosis; in the histopathological

findings: a diffuse gliosis degenerating into a glioblastoma multiforme; and in the light cast on the structural basis of emotion. The case is like an experiment designed by nature to support Papez's view of the mechanism of emotions.

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