APPercPtiVe BlinDNESS IN LISSAUER'S DEMENTIA Paralytica

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Pick (1898) described as apperceptive blindness the inability to see resulting from loss of visual attention. He observed that symptom in cases of circumscribed atrophy of the occipital lobes. According to Pick the loss of visual attention resulted in an inability to focus and in a loss of visual apperception, i.e. of the capacity for conscious perception of visual stimuli. Pötzl (1928) who studied the symptom in the same kind of case material expressed the opinion that apperceptive blindness belonged to the visual agnosias, the basic disorder being an inability to synthesize visual impressions. Observations made by Horn and Stengel (1930) in a case of Pick's disease corroborated Pötzl's view. The literature does not contain further reports on that symptom which obviously occurs only under a specific set of conditions of which little is known. We have observed a patient who to our knowledge is the first reported case of apperceptive blindness in general paralysis; it allows us to carry the analysis of that symptom further than has been possible hitherto.

Mrs. C. M., aged 27 admitted 16/2/42. Five weeks prior to admission she had been delivered of a living child. After delivery she was delirious for three days and during the following weeks she was strange and noisy. On admission she gave the following history of herself. When she was five her mother died. The father, an alcoholic, committed suicide in 1930. The patient did well at school. For three years she was cared for in an orphanage which she left at the age of 15. The only facts that could be obtained about the following years were that she lost two domestic posts through pilfering and was treated for syphilis. She married at 20. The husband is healthy. The patient has had four healthy children and one still-birth. During each pregnancy she received anti-syphilitic treatment but was a chronic defaulter.

Condition on admission.—Patient was over-talkative and emotionally unstable. Questions were answered promptly, sometimes in a rambling manner. She made grammatical mistakes, especially in the use of pronouns; she would use "she" instead of "he," "I" instead of "she," etc. (Who was Napoleon? "She wanted to conquer the world." (How is your husband?) "I have a letter from her. I am a prisoner in Germany." Except for those grammatical mistakes there were no signs of aphasia. The speech was slightly monotonous, but there was no marked dysarthria. The orientation was correct. Memory and retention were considerably impaired. Her general knowledge was poor. She could not do simple multiplication and subtraction was slow and inaccurate. The patient lacked real insight.

Physical examination.—Physical condition satisfactory. No stigmata of congenital syphilis. Cardiovascular system normal. Pupils regular, reacting briskly to light and accommodation. Fundi healthy. No signs of hemianopia. Neurological examination negative.—WR in blood positive. CSF: WR strong positive, lymphocytes 10, globulin increased, Lange 5555554310; Chlorides 70; Glucose 58.

15/3/42. The behaviour of the patient was satisfactory and she was allowed to go out unaccompanied. 2/4/42. Status epilepticus lasting for one-and-a-half hours, controlled by intravenous phenobarbitone sodium injection. 4/4/42. The patient, who had been drowsy since the status epilepticus was slightly confused and when asked to name objects she repeatedly said she could not see. She obviously did not focus. She could, however, on several occasions name objects correctly without the help of touch or sound, thus proving that she was not blind; on superficial observation she sometimes gave that impression as she often knocked into objects. In her speech she showed a severe degree of perseveration. 12/4/42. Delirious with visual hallucinations. Saw her husband and children outside and talked and waved to them; occasionally she seemed to hear their voices. Did not recognize anybody in the ward. Did not fix her eyes when requested to look at objects. Repeatedly asserted that she could not see or that she could not see well. 13/5/42. Now recognizes doctor and nurses. Expressing vague paranoid ideas. 20/5/42. Produced visual hallucinations and was inclined to confabulate. Holding her bedspread as newspaper she read: "Tonight I will be killed with an injection. I'll be ending my life on the scaffold." 24/6/42. At times distressed and delirious. She still asserted she could not see and at the same time expressed visual hallucinations in which she saw her children and talked to them.

29/7/42. The delirious condition had subsided and the patient was more co-operative. She could not for any length of time focus her attention on a task. In conversation it often was impossible to overcome the severe perseveration. Frequent response to any question she found difficult to answer was the utterance "I'm stumped." She would only very rarely say "I don't
know." Often she talked away in a confabulatory manner. When shown objects she focussed only occasionally and then would give the correct answer, sometimes requesting to be allowed to touch the object, but sometimes (especially when asked to name something) did not focus at all. The extreme lack of attention to visual stimuli was also shown by the fact that the blink reflexes were often missing on both sides. Another response was a perseveration of naming some object with which something happened to be in her visual field, usually connected with the body of the examiner, instead of the object shown to her. It appeared that the patient, instead of focussing on the object presented picked out some part of the background. On other occasions she named details of an object but was unable to comprehend the whole. Her behaviour and her utterances gave rise to the impression that the visible world presented a chaotic picture to her. She tried easily and then the perseveration and the alleged inability to see became more marked. Right and left were often mixed up and when asked to name her own or the examiner's fingers she usually said she could not see, or used some other stereotyped excuse.

Her speech was slow and somewhat dulling. There was no dysarthria and she was very talkative especially when spoken to. She would attract her attention and understand spoken language well. She occasionally produced wrong words as the result of perseveration. It was difficult to make her read and write. She usually said she could not see and made no effort to comply with requests to write. Once one object had been named, the numbers correct but it was never possible to make her calculate. Her writing was disturbed. She did not stick to the line. Asked to write her name she wrote " M ", then stopped and wrote " rs " in the line above. Then she proceeded to write the name (Murphy), writing " M " with an extra loop followed by " ur " then another " r " higher on the paper covered by a large and distorted loop of an incomplete " p " . Asked to write " hospital " she wrote " hosperl " and " hospeel " . Other specimens showed a similar inability to keep to the line and the tendency to distort words by perseveration and omission. When given print to read she occasionally read a few words correctly, never a full sentence; usually she pushed the paper aside saying she could not see. When shown pictures she responded in the same way. It was impossible to persuade her to try drawing or copying. There were no signs of apraxia; she took her food without difficulty and handled objects of everyday use correctly; it was not possible to test her for any other functional apraxia.

The normal withdrawal or blinking reactions to strong flashes of light suddenly brought into her visual fields, were either lacking or very deficient; nor did she withdraw when an open knife was suddenly brought near her face. The same lack of response was noted when sudden loud noises sounded near her ear; she did not turn reflexly to the source of the noise nor did she withdraw her head. The reaction to pain was abnormal. There was no reaction of withdrawal when her hands or any other part of her body were strongly pricked with a pin; when the outstretched tongue was pricked she did not withdraw it either. There was on the whole a lack of responses to outside stimuli, however painful and apparently dangerous.

10/9/42. Status epilepticus after which a complete right-sided spastic hemiplegia was found. 26/9/42. During the days following the epileptic state the patient was drowsy and later became delirious. She rapidly went downhill and died with signs of broncho-pneumonia and cerebral oedema.

Post Mortem examination.—(Dr. W. Blackwood). Basal congestion in both lungs. Brown atrophy of the heart. Two small recent infarctions in the left kidney. The microscopic examination was in accordance with the naked-eye appearance. Other organs healthy.

Dr. E. Stengel. Generalized symmetrical gyral atrophy, moderate in the frontal and temporal regions, more marked in the posterior parts, especially in the occipital lobes. Opacity of the leptomeninges, corresponding to a chronic diffuse leptomeningitis. Moderate hydrocephalus internus and dilatation of the 3rd and 4th ventricle.

Microscopic examination. Lepto-meninges.—Increased in fibrous tissue; number of blood vessels increased, many of which showed infiltrations consisting of plasma cells and lymphocytes. The inflammatory changes were much more diffuse and cortical parts of the cortex, especially the area parastriata. There were deposits of haemosiderin-like pigment in adventitial and microglial cells. Some of those deposits gave a positive iron reaction. The cytarchitecture of the cortex showed considerable pathological changes. There was a marked loss of ganglion cells, particularly in the 3rd and 5th layers, and many pyramidal cells showed disorientation. All phases of degenerative cell disintegration could be demonstrated. Those changes were again most marked in the occipital region, but the area striata was comparatively less affected than the adjoining areas. The loss of nerve cells in the anterior central gyrus was comparatively moderate and no circumscribed lesion could be found. There was a considerable degree of diffuse demyelinization in the cortex which reached its maximum in the parieto-occipital areas; there were the usual signs of lipoidal breakdown, again more marked in the posterior cortical areas. Diffuse microglial proliferation over the whole cortex, in accordance with the local degree of the pyramidal process. In the cornu Ammonis the changes were considerable and of the same nature as in other parts of the cortex. Small subcortical patches of demyelinization were found in the occipital and frontal areas. Moderate degree of inflammation in the region of the ganglia, particularly in the putamen. In the cerebellum some arteries showed cuffing. Sections of the medulla and the spinal cord did not show pathological changes.

Summary
This case of general paralysis came under observation following the birth of her fifth child which had precipitated a delirious condition. Three months later the patient had a status epilepticus which was followed by the appearance of atypical symptoms of a localizing nature. The patient maintained that she could not see. This statement alternated with vivid hallucinations, chiefly visual in character. Later the hallucinations subsided and the patient exhibited a complex clinical picture among the components of which the symptom of apperceptive blindness was prominent. The inability to focus was very marked. Though the general mental condition of the patient made a systematic examination difficult the observations collected were sufficient for ascertaining that there were symptoms of a visual agnosia, a disturbance of right-left orientation and most probably a finger agnosia with agraphia. The writing suggested that one of the components of the agraphia was a disturbance of spatial orientation. There was perseveration in speech and actions and a tendency to dodge more complicated tasks. The understanding of spoken language was not impaired. The response to pain and other external stimuli corresponded to what Schilder and Stengel (1930) have described as "asymbolia for pain." The patient died following two attacks of status epilepticus; after the first attack a hemiplegia had developed.

The post-mortem examination revealed changes
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characteristic of a general paralysis. Those changes were much more marked in the occipital area, than in other parts of the cortex. Within the more heavily affected areas there were again some quantitative differences, the area striata being less affected than the other parts of the occipital cortex. Nowhere were there changes of the type described as status spongiosus. The changes suggested an acute exacerbation of a typical paralytic process exhibiting both the inflammatory and the degenerative features.

Comment

In this case the diagnosis of Lissauer's dementia paralytica was justified on the ground that there were localizing symptoms pointing to a comparatively more severe affection of the parieto-occipital cortical areas. The occurrence of epileptic seizures and of a hemiplegia supported that diagnosis. Alzheimer (1904) regarded the "posterior" distribution as characteristic of the Lissauer type of general paralysis. It is not intended to enter into a discussion of the pathology of Lissauer's dementia paralytica re-examined recently by Galbraith and Meyer (1942) who described a case in which the character and distribution of the lesions were the same as in our case. Our findings confirmed the conclusions of those authors that the pathological process underlying Lissauer's dementia paralytica is not necessarily a status spongiosus as generally assumed. Galbraith and Meyer regard their case as a fairly recent one but they were unable to trace any precipitating factor. It is noteworthy that in our case the acute exacerbation of the paralytic process resulting in the picture of Lissauer's dementia paralytica developed soon after a delirious condition following childbirth. This is of interest in view of the opinion expressed by neuropathologists that the Lissauer's type of general paralysis is due to a toxic factor superimposed on the general paralytic process.

The most interesting feature of this case was the symptom of apperceptive blindness. In trying to analyse that symptom it is necessary to consider the whole of the patient's mental condition. There was, apart from the dementia, a severe thought disorder of a type which often follows frequent major epileptic attacks or head injuries (Symonds, 1937, A. Paterson, 1942). Retardation of all mental activities with perseveration was an expression of that thought disorder. It is significant that apperceptive blindness appeared after the first status epilepticus. There were in addition symptoms of a visual agnosia. In attempting to comprehend the outer world visually the patient either failed completely or sometimes set about that task in a piecemeal way, picking out details and thus trying to synthesize the visual impressions. Certain responses could be best described as the result of an inability to separate "figure" and "background" (Gelb and Goldstein, 1918). Another feature of the patient's performance was that occasionally she would recognize and name an object correctly in its natural setting whereas she would fail to do so when the object appeared in the "artificial" situation of the examination. Her behaviour was in accordance with that described by Gelb and Goldstein (1918) in their case of visual agnosia. However, it is clear that our case cannot be regarded as proof of the correctness or falsity of the Gestalt theory of pure visual agnosia. The experience of not being able to see appears to be the result of the general disorder of mental activity, as expressed by thought disorder, lack of attention and inability to focus, interacting with a visual agnosia. Those disorders apparently resulted in the visual impression of a chaotic outer world, which, as it were, blinded the patient; she could not but describe that experience as an inability to see. It is noteworthy that the patient never described herself as blind. There obviously is a great variety of sensations which result in the experience of an inability to see. It is well to remember that even the way in which hemianopia due to cerebral lesions is experienced differs in various individuals, some experiencing the lost part of the visual field as a dark gap ("vision noire" of French authors), others perceiving only the remaining visual field ("vision nulle"). It appears that even in those well-defined lesions the individual reactions of the patients determine the subjective effect. To describe the symptom of apperceptive blindness as a reaction rather than an effect is justified on the basis of the observations that the same individual would sometimes attempt to comprehend the outer world in the same way as a patient with pure visual agnosia. Patients such as ours are at a serious disadvantage compared with those suffering from pure visual agnosia. The general mental disorder makes it impossible or much more difficult for them to "circumvent" their defect. The question arises whether that symptom should not be regarded as simply hysterical. However, such a description would be misleading. Present-day usage of the term hysterical for a symptom occurring in organic cases implies a hysterical feature additional to the organic picture and is supposed to permit of deductions as to the premorbid personality. An analysis of apperceptive blindness on such lines would not only lack the clinical basis but it would not do justice to the fact that similar reactions structurally similar can be observed in many cases of brain disease. Hughlings Jackson has shown that many of the responses of such patients are to be understood as reactions to their disabilities. Goldstein (1939) has explained many features of the behaviour in organic as well as psychoneurotic disorder as the expression of a tendency to avoid catastrophic situations, i.e. complete breakdowns of mental activities. Our patient was, as the result of her general mental involvement, unable to cope with the difficulties imposed on her by the localized cortical lesions. In her relation to the outer world a catastrophic situation threatened to arise. The assertion that she was unable to see can be understood as a denial of the chaotic condition of the
outer world. Thus apperceptive blindness can be described as a primitive way of avoiding a catastrophic situation by denial. The similarity of this mechanism with those underlying certain psycho-neurotic reactions is obvious.

There are some other observations suggesting that the adjustment of the individual to changes in the visual aspect of the outer world is sometimes particularly difficult, especially when those changes arise fairly suddenly and when in addition to a disorder of the visual functions there exists a gross general mental disorder. Then the mechanism of denial sometimes intervenes resulting in such symptoms as the unawareness of cortical blindness (Anton, 1899). That symptom which is usually associated with visual hallucinations of a confabulatory character, though apparently the opposite of apperceptive blindness, is psychopathologically akin to it; the mechanism underlying both symptoms is that of a denial of a complete change of the visual aspect of the outer world. In this connection it is of interest that our patient had for a time visual hallucinations of exactly the same type as occur in Anton's symptom. Those hallucinations can be understood in our case as another attempt at a denial of the chaotic condition in which the outer world presented itself. In both apperceptive blindness and Anton's symptom an awareness of the disability occasionally breaks through. In the analysis of Anton's symptom, which is an instance of anosognosia (Babinski, 1914), the relationship to the hysterical phenomena was noted and the mechanism of denial of organically produced defects was described as "organic repression." Another instance of anosognosia affecting the visual apparatus is the symptom of imperception for the position of the eyelids recently described in this journal (Rubinstein, 1941). All those observations show how much the reactions of the individual in brain disease have in common with those in psychoneuroses. This similarity suggests that both groups of mechanisms follow common biological laws.

Conclusions

A case of Lissauer's dementia paralytica with apperceptive blindness has been described. The pathological examination revealed the typical changes of general paralysis which were more marked in the posterior parts of the cortex and reached their maximum in the parastral areas.

The symptom of apperceptive blindness which hitherto has been observed only in cases of Pick's disease with occipital involvement has been analysed. Apperceptive blindness has been described as a reaction to a chaotic visual impression of the outer world as it presents itself to patients with a severe general mental disorder interacting with a visual agnosia. The main features of the mental disorder were an organic dementia and a thought disorder of the type seen in chronic epilepsy with frequent fits. It has been shown that the psychological mechanism underlying apperceptive blindness is that of denial and it has been pointed out that apperceptive blindness is genetically akin to the symptom of unawareness of cerebral blindness and other instances of anosognosia. The relationship of that group of reactions in brain diseases to psycho-neurotic symptoms has been discussed.

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