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COMPULSIVE FEATURES IN A PATIENT WITH LEBER'S DISEASE

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ALTHOUGH reports of cases of Leber's disease (hereditary optic atrophy) have been published by British, French, and German authors, very few such reports are to be found in the American literature. The unusual findings of neurotic manifestations of a compulsive nature in a female patient with this disease appears to justify their presentation.

Julia Bell (1931) has made an extensive study of the literature on the subject of Leber's disease and quotes the works of Beer (1817), Von Graefe (1858), and Leber (1871). Leber's disease usually begins at puberty or in early youth and is characterized by a rapid loss of vision, contraction of the visual fields, central scotomata and primary optic atrophy. Slight improvement may follow the primary loss of vision.

Case Report

E.B., a 32-year-old married Jewish housewife, was admitted to the Bellevue Psychiatric Hospital on 5th January, 1936. She had been referred for mental observation from the Bellevue Mental Hygiene Clinic, as she was said to have a compulsive neurosis, optic atrophy, and suicidal tendencies. She was born in New York City of Russian Jewish parents, and graduated from a grammar school at the age of fifteen years. At that time on the Stanford Binet tests she was rated as of dull normal intelligence with a mental age of twelve years and two months. Except for doing office work for a few months, she remained at home caring for her mother until the time of her marriage at the age of nineteen.

The past history was essentially negative except for diphtheria at the age of nine

or ten. The birth and early developmental history were negative. There was no history of convulsions nor of neurotic traits in childhood. Her menstrual periods had been regular since their onset at the age of fifteen years.

The patient's eye symptoms began suddenly in July, 1923, at the age of nineteen, one month after her marriage, when she suddenly lost her vision almost completely and it was noted she developed strabismus. The patient had been having a few headaches previously and had a "cold." The patient said that she went to bed one night feeling well. On waking the following morning her vision was impaired. Everything appeared "in a fog." During the course of the day the vision became progressively worse and continued to do so for three or four weeks, when some slight improvement set in. During this period the strabismus developed. A year after the onset some further slight improvement followed an operation on a nasal sinus.

The patient had been under the care of Dr. Mark J. Schoenberg intermittently since 1924. He reported that in 1924, when he first examined her, she had bilateral optic atrophy of several months' duration and her acuity of vision was reduced to counting fingers at two feet in each eye. In 1934 the visual fields showed a binasal defect, almost hemianopic, and included the central portion of the fields. In September, 1935, the vision was 1/70 (O.D.) and 1/200 (O.S.). There was a central opacity of the lens in the right eye. No mention was made of the existence of the cataract in 1924 or 1934. Apart from the "cold" there was nothing to suggest a "toxic" basis for the eye disturbance. Signs of increased intracranial pressure were also absent.

The neurotic symptoms began about July, 1934. The patient described her symptoms as follows: "I'm nervous about a year and a half. At the beginning I used to linger over things. If I did something I would stand over it and see to it that it wasn't wrong. I would try to make sure. I would repeat it over and over. I did the same thing three or four times. I can't control myself. I mumble to myself and talk to myself. I think too much of myself . . . I started touching my nose and eyes, and then I would see if the floor was clean and I would go over that three or four times. Then I would wipe the dishes and I would wipe them over and over again. I then would see if my clothes were fixed straight and I'd pull and straighten them over and over. Then I'd look to see if anything was lying around and I'd go back to be sure . . . I wasn't sure of my eyes so I would go back and do the same things over and over again. I couldn't see very well and I wanted things to be clean, so I would dust and dust over and over to make sure. . . . I'm disgusted with life and want to die. I get practically no enjoyment from sex life. I'm still afraid to get pregnant. I'm not a fit person to have children. If I had money I'd get an operation just to be sure I'd never get pregnant."

Examination.—The general physical examination was essentially negative except for the signs of pregnancy of three months' duration. Smell was normal. There was a divergent squint. The right pupil was slightly larger than the left; both reacted promptly to light and on accommodation. There was a right posterior polar cataract and bilateral primary optic atrophy. Visual field examination showed slight nasal constriction in the left eye, but these tests were unsatisfactory owing to the large size of test objects necessary. A definite central scotoma was present on the left side. The patient was colour-blind. The right iris had less pigment than the left. There was no nystagmus. Corneal and nasal reflexes were active and equal. Hearing was normal. The deep and superficial reflexes were normal. There were no signs of ataxia or dysmetria, nor any loss of associated movements in walking. X-ray and laboratory findings, including blood, spinal fluid, and urine studies, were essentially negative.

Behaviour under Observation.—At the time of her admission she was tense, restless, agitated, and admitted suicidal tendencies. She showed constant repetition of certain

movements, such as tugging at her clothes, rubbing her eyes, or picking her nose. She would repeatedly grunt and mumble to herself. She would cry frequently. At times she showed choreiform movements. She had difficulty in sleeping at first and required sedatives. After a few days she began to improve and gradually she had fewer and fewer compulsions. The psychotherapy given her consisted mainly in suggestion and reassurance, together with assistance in altering the environmental factors, particularly with regard to the amount of housework she had to do. She was referred to a birth-control clinic for contraceptive advice. She was discharged to her husband's custody on 18th January, 1936, and then was followed in the Mental Hygiene Clinic. In June she gave birth to a normal full-term baby. She has continued to show improvement.

When she was examined on September 4th, 1936, she said: "I had a baby three months ago. Now I can do some of my housework. I can take care of my baby, dress, and feed him. I do the shopping and the cooking. Before, I didn't have the patience or desire to do any of the work. Now the day goes by fast and I sleep well. I've made up my mind that nothing is going to bother me. I no longer moan and groan. Once in a while I catch myself beginning to do things, as straightening something out. Then I make up my mind and it doesn't bother me."

Family History.—The patient is the fourth oldest of five siblings. The father, aged sixty-eight, is still alive. He works as a presser in a shop. He has had no eye symptoms. The mother was an invalid for seventeen years, prior to her death in 1935. She apparently had a cerebral accident with hemiplegia. The oldest sibling has similar eye trouble. Two other brothers and a sister are free of eye trouble and of any other neurological or psychiatric disorders. Two maternal male cousins were described as having "eye trouble" similar to those of the patient and her brother. Their visual disturbances have been present for many years. No details were elicited and the men were not available for examination.

The patient's brother, B.R., aged thirty-eight years, had a sudden loss of vision in 1929 at the age of thirty-one. He said that he lost his vision suddenly. He observed that his eyesight, previously excellent, suddenly became blurred at the movies. On leaving the movies he mentioned that he suffered from misty vision and was unable to read. These symptoms lasted for fourteen days, and then improved a little. For one and a half years he could not recognize colours, but improvement has taken place. Physical and neurological examinations were completely negative, except in the eyes. There was bilateral primary optic atrophy; the discs were more blue than those of his sister. There was bilateral concentric constriction of the visual fields. No central scotomata were present, although there was a relative scotoma for colour bilaterally. At a distance of 1 metre he was unable, using his left eye, to name various coloured test objects accurately if they were less than 6 mm. in diameter. With the right eye he could not detect coloured objects less than 2 cm. in size. A few nystagmoid jerks could be elicited. The patient had weakness of ocular convergence. The pupils were slightly irregular, but reacted promptly. The remaining cranial nerves were negative. The deep and superficial reflexes were intact. Laboratory studies were negative. This patient had no neurotic symptoms. His sexual power was intact. He was employed as a plumber's helper and as a petrol station attendant prior to his illness. He appeared to be of average intelligence.

Discussion

Although it is a well-accepted fact that neurotic symptoms may occur in organic diseases of the nervous system, I have been unable to find case reports of obsessive or compulsive neurotic symptoms in persons with Leber's disease.

Schilder (1929), in his article on "The Somatic Basis of the Neurosis," says : ". . . of course every neurosis must have an organic basis, because normal life, normal psychic life, also has an organic background and one cannot make a distinction between the organic and the psychic." Jelliffe (1932) and Schilder (1931) describe compulsive and obsessive features in post-encephalitic patients and discuss regressive tendencies resulting from the disease. Jelliffe describes one post-encephalitic patient who had the compulsion to count letters and another who had a compulsion to think of dirty things. He also emphasizes the symptom of anxiety in his cases. Bender (1935) describes a patient with compulsive thoughts of a sexual nature who had an obscure encephalitic-like illness three years earlier and had some neurological signs, including tremors, flattening of facies, loss of associated movements, and hyper-reflexia. Schilder, in a personal communication, describes a patient he once treated who had bilateral congenital cataract and who, after several operations, developed obsessional trends in connection with his defective vision in which he worried if there were not a spot on his tie or whether he had enough of a shave.

Stengel (1935) in a recent article summarizes most of the literature dealing with the problems of obsessive and compulsive phenomena in organic brain diseases from a psychoanalytical viewpoint. He discusses disturbances of the Ego in cases of epilepsy, in hemichorea following encephalitis, and in oculogyric crises in Parkinsonism. He describes one epileptic patient who had the compulsive idea concerning why the letter "o" should be round, how letter-writing originated, and what would happen if the patient were three thousand years old. Other patients showed compulsive thoughts with open sadistic and masochistic ideas, such as dealing with the thought of killing the father, etc. He also emphasizes the work of Stern in connection with oculogyric crises (Jelliffe, 1932), and the works of Burger and Mayer-Gross (1928). Stengel elaborates the idea that the patient's Ego satisfactions are limited by organic brain disease which impairs the normal sensory or motility functions. He infers that the compulsive or other neurotic symptoms are based on this mechanism rather than on the structural organic defect.

It is worthy of note here that our patient describes herself as always being unusually neat, clean, orderly, always wanting things properly arranged, etc. As a result of her visual impairment she can no longer verify the neatness or orderliness of her appearance, her clothes, or her household goods. As a result she begins to repeat certain rituals, such as the arrangement of her dress, the dusting of a chair or table until a neurotic syndrome develops in which she must repeat these acts over and over. The fact that her symptoms receded quite rapidly under moderate psychotherapy tends, in my opinion, to indicate a more favourable prognosis here than in the usual neuroses growing out of the infantile situation. The prognosis for the neurosis arising in patients with post-encephalitis has been shown by Bender to be less amenable to psychotherapy than the usual neuroses ; this would not, however, apply to neuroses associated with other somatic diseases.

It must be recognized that the relationship between the organic disease and the neurotic syndrome in this woman is probably not the same as that between

the organic brain disease and the neurotic symptoms in the cases described by Jelliffe, Schilder, Bender, and Stengel, although Jelliffe does make the point that the neurotic behaviour is an expression of the individual's (Ego's) inability to cope with the insult to the personality as a whole. It is probably fair to assume that there is no definite organic relation between the ocular pathology and the neurotic picture in this woman unless one assumes that the compulsive syndrome is an evidence of neuropathic weakness in a woman who has a constitutional disease in another part of the nervous system. It is easier to understand her symptoms in the light of her emotional conflicts. It cannot be insignificant that the compulsive, obsessional, and depressive features of her illness reached their height in the third month of pregnancy. Other points which are significant are that her husband states that she is frigid and often refuses sex relations and she has induced two miscarriages. The beginning and the progress of her symptoms are best understood in the light of her disease and the handicap it forces upon her and her resulting struggle to fulfil her duties as a wife and mother. The compulsions start with the tendency to touch her eyes and nose. She then has the compulsion to clean things over and over because she cannot trust her eyes. Then, because of her poor vision, she never has the satisfaction of seeing anything looking clean and immaculate. Everything always looks dirty. Even now that she is essentially free of her compulsions she says that it is the spots before her eyes that make everything look dirty. Then she works from seven in the morning to midnight trying to do her duty by her home, her children, her husband, and her sick mother. She is never satisfied. It is likely that a deep-seated feeling of inadequacy and guilt forces her to such work. We can postulate that this woman sees herself faced with three alternatives. She must refuse sex relations to her husband and fail as a wife, and we do not know with what grace the husband accepts this ; or she must see herself develop a pregnancy of which she is in constant dread ; or she must induce abortions, as she has done twice already. She was persuaded to enter the hospital at the time she contemplated suicide in the third month of pregnancy and there became reconciled to the pregnancy ; this was probably an important factor in the good results from the psychotherapy that was given her. The conflicts are temporarily resolved, although from time to time even yet she recognizes the slight tendency for the familiar compulsions to recur. Her brother is happily saved from these many conflicts.

Summary

A case is described of a female patient having the classical symptoms of Leber's disease. In addition, she had symptoms of a compulsive neurosis, and these symptoms have diminished under psychotherapy.

REFERENCES

- Bell, J. (1931). *Treasury of Human Inheritance. Anomalies and Diseases of the Eye.* IV. *Hereditary Optic Atrophy (Leber's Disease).* Cambridge University Press, London.
- Beer, G. J. (1817). *Lehre von den Augenkrankheiten*, 2, 443, Wien.
- Bender, L. (1935). *Amer. J. Psychiat.*, **92**, 325.
- Bürger, H., and Mayer-Gross, W. (1928). *Z. ges. Neurol. Psychiat.*, **116**, 645.
- von Graefe, A. (1858). *Arch. Ophthal.*, **4**, 266.
- Jelliffe, S. E. (1932). *J. nerv. ment. Dis. Monograph*, Series 55.
- Leber, T. (1871). *Arch. Ophthal.*, **17**, 249.
- Schilder, P. (1929). *J. nerv. ment. Dis.*, **70**, 502.
- (1931). *J. nerv. ment. Dis. Monograph*, Series 53.
- Stengel, E. (1935). *Internat. Z. Psychoanal.*, **21**, 544.