A Unique Illness Involving the Cerebral Cortex

A case report from the Mental Institution in Frankfurt am Main.*

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The clinical course and pathology of this distinctive process separate it from the known neurologic disorders.

The patient initially presented at fifty-one years of age with jealousy of her husband. Rapidly progressive memory loss soon followed. No longer finding her apartment suitable, she dragged her furniture back and forth and concealed it. She began to believe that others wanted to kill her, and she would scream out loud.

Following institutionalization she appeared totally bewildered. She was disoriented as to time and place and occasionally stated that she did not understand events around her. She treated her physician as a guest, excused herself and said she was not finished with her work. Following this she would scream aloud that he was trying to stab her with a knife, or indignantly turn him away, fearing that he would violate her. She was intermittently delirious, dragged her bedding about, called for her

*The clinical examination and central nervous system autopsy were performed by Dr. Sioli, director of the institute.

husband and daughter, and appeared to be having auditory hallucinations. She would scream for hours in a monstrous voice.

She was unable to understand situations and would scream every time someone would attempt to examine her. Only after repeated attempts could an examination be performed.

When shown objects she could name them relatively correctly. However her perceptions were extremely disturbed. Immediately after naming the objects she would forget them. She drifted from one line to the next while reading—either enunciating the individual letters or speaking in a meaningless tone. While writing, she repeated single syllables, omitted others, and quickly became confused. She used perplexing phrases when speaking or made paraphasic errors ("milk pourer" instead of "cup"). She would hesitate during speech. She did not understand some questions put to her. She appeared to have forgotten the use of several objects. Hand functions and walking were undisturbed. Her patellar reflexes and pupillary reactions were preserved. Her radial arteries were rigid. There was no cardiac enlargement or urinary albumin.

Her focal symptoms clearly waxed and waned; but they remained minimal during her illness. However her mental deterioration was progressive. She died after four and a half years of illness. Prior to her death she had become completely apathetic, went to bed in her clothes, neglected her personal hygiene, and developed decubitus ulcers despite nursing care.

The brain sections demonstrated generalized atrophic changes. The major cerebral vessels showed atherosclerotic changes. Neurofibrillary changes were seen in sections stained by the Bielschowsky silver technique. Thick, heavily stained fibrils stood out among the few remaining normal cells. There were many similar fibrillary networks which merged into dense bundles and finally appeared on the cells' surface. In the most advanced stage the cell nucleus and cell body disintegrated leaving only a tangled bundle of fibrils in the site of a former ganglion cell.

These fibrils stained differently from normal fibrils, suggesting that a chemical transformation of the fibrillary substance must have occurred. This transformation may explain the preservation of fibrils despite the cellular destruction. The alteration of the fibrils appears to go hand in hand with the deposit of a previously undescribed material in the ganglion cells. One quarter to one third of the cortical ganglion cells, particularly of the superficial cell layers, had disappeared.

The entire cortex, especially the superficial portions, showed miliary foci resulting from the deposit of a unique substance. This material could be recognized by virtue of its refractivity on unstained sections.

The glia showed numerous filaments and fatty vacuoles. There was no vascular infiltration; but endothelial proliferation and neovascularization were seen.
auditory hallucinations.

would scream every day after repeated attempts were made to wake her up. Immediately after naming numbers, odd or even, she used perplexing phrases like "milk pouer" instead of the correct term. Her speech did not understand or repeat the use of names. Her radial artery and urinary albumin were normal but they remained uncontrolled. Her speech showed a deterioration was progresses. Prior to her death, she was in her clothes, with multiple ulcers despite trophic changes. The skin showed fibrinous silver staining, the few remaining nerve cells which merged at the surface. In the disintegrated nerve cell bodies, silver staining was present. No fibrils were seen despite the use of silver impregnation technique. This material could be seen in sections.

We feel that this represents a unique entity. In the last few years, more cases have been confirmed. These observations suggest that we should not be content to classify a clinical case without exerting maximal effort to investigate it. There are without a doubt many more illnesses than our textbooks describe. In some instances the peculiar features will be confirmed histologically. Over a period of time we will come to the point where we can isolate single clinical cases from the larger classifications and thus more clearly define each clinical entity.