



Historical Clinicopathological Conference

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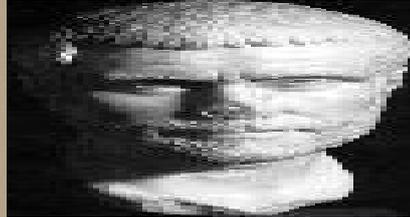
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2001

PRESENTATION OF CASE



In the month of October, a 63-year old man developed postprandial abdominal pain and vomiting. He had been feeling well until passing out during a banquet in which he had consumed a large quantity of wine and a variety of dishes, of which one composed of mushrooms was a long-time favorite. On regaining consciousness a short time later, he complained of severe abdominal pain. He vomited and felt somewhat

better.

Prior to this illness, the patient had enjoyed reasonably good health for over a decade. However, early in life he had been sickly. He was born prematurely (after seven months gestation) and as a child, suffered from a succession of obstinate disorders, including milk allergy and frequent febrile illness. He is also believed to have had malaria, measles (complicated by unilateral deafness), erysipelas and colitis. Since early childhood, he had weakness of both legs so pronounced that he limped noticeably and could ambulate no further than short distances without assistance. He had long-standing, intention tics and jerks of his head and hands, as well as stammering and drooling, which were most pronounced when he was excited. He was also prone to fits of inappropriate laughter. He regularly ate and drank to excess, rarely leaving his dining room until he was both "stuffed and soaked." Thus, it was not unusual for him to fall asleep immediately after dining and to have to be carried unconscious to his bed. He had gained considerable weight in recent years and had been complaining bitterly of episodes of heartburn so frequent and severe that he contemplated suicide as a means of relief.

The patient's father died of trauma at age 28; his mother committed suicide at age 73. He had one brother who had died at age 34 of a mysterious illness manifested by progressive wasting with terminal acrocyanosis and a trunkal rash. His sister was executed for adultery at age 34. There was no family history of neurological abnormalities similar to the patient's.

The patient was born in France, but had spent most of his life in Italy. He had been married four times and had also had numerous heterosexual relationships, including several with prostitutes. He had three children, all of whom were alive and well at the time of his illness. He was a politician and a historian.

Physical examination showed an obese man in moderate dress. The temperature was normal. The eyes were injected. The hearing from impaired unilaterally. The abdomen was mildly tender throughout. The voice was hoarse and indistinct. The patient stammered uncontrollably in response to questioning with considerable slobbering and rhinorrhea. The neck muscles were enlarged. The muscles of his upper extremities were well developed and strong, whereas those of both lower extremities, especially the calves, were weak and atrophied. When the patient walked, he dragged his right foot.

A physician induced additional vomiting by placing a feather in the back of the patient's throat. Shortly thereafter the patient's condition deteriorated. He became confused and exhibited signs of unremitting abdominal pain and fecal incontinence. He expired 12 hours after the onset of his illness.

[Click here for final diagnosis](#)