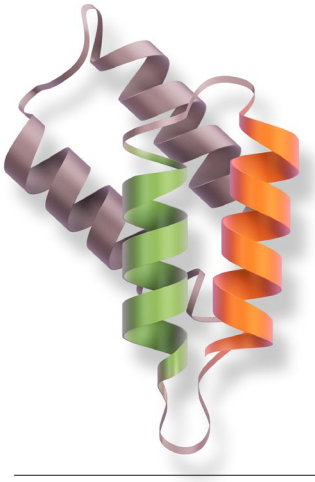

NATIONAL CENTER FOR CASE STUDY TEACHING IN SCIENCE

Tremors in the Nerves: Investigating Kuru

by

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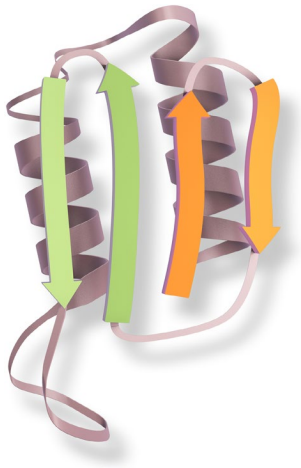

You are on a team of administrative physicians tasked to provide oversight of diagnostic care for your local hospital. Your team has just obtained a hypothetical case on Patient SG, a 60-year-old nurse who worked in Papua New Guinea early in her career where she was exposed to many sick patients on a daily basis. Due to limited supplies, Patient SG had to administer stitches to patients without wearing gloves. While her memory of the situation is not clear, she believes that she likely came in contact with patient blood on a regular basis. Later in life, she noticed that she started to have a tough time holding her medical instruments, exhibited symptoms of persistent bodily shaking and could not control her muscles to stop it. As time progressed, she started having difficulty walking and began to develop depression-like symptoms. Furthermore, her colleagues noticed that she started to have a tough time talking and suggested that she visit a doctor.

Patient SG had returned to the United States years ago, and thus went to the local hospital in her hometown. She reported in her medical history that she had been healthy her entire life, with exception to a stroke she had ten years ago. She insisted that the stroke was stress related. Based on presented symptoms and medical history, the hospital suspected that she most likely had Parkinson's disease, a neurological degenerative disease that affects the motor function in the central nervous system. The typical symptoms associated with Parkinson's disease are shaking, slow movement, depression-like symptoms, increased risk of stroke and issues with walking. Parkinson's disease is caused by death of dopamine neurons in the brain and it is not associated with the prions that cause Kuru. There are currently no specific tests to definitively diagnose Parkinson's disease. Furthermore, there is no cure for the disease; medications are given to control symptoms related to walking.

You receive the blood test, EEG, and MRI results. A traditional comprehensive blood test reports no irregularities in the blood. The EEG shows that there are disruptions in neurological functions and the MRI shows that there are structural damages in the brain, specifically lesions in the cerebellum. The patient was prescribed with medications to combat Parkinson's disease but they proved to be ineffective, since Parkinson's disease occurs in a different part of the brain called the basal ganglia. The doctors assumed the lesions in her cerebellum were stroke related. Her symptoms worsened and she became unable to walk at all, confining her to a wheelchair. Later that week, she passed away.

An autopsy was conducted and because of her early nursing career work with tribes in Papua New Guinea, a pathologist decided to test for Kuru because of the unique protein aggregate distribution throughout her cerebellum. A test of Patient SG's cerebrospinal fluid revealed infectious prions postmortem. This disproved the initial diagnosis of Parkinson's disease and suggested that the patient had most likely obtained Kuru disease early in her life. The progression of Kuru can take decades and symptoms may not present for many years.

Questions

1. Kuru disease is commonly contracted through the cannibalistic rituals of the Fore people of Papua New Guinea. However, Patient SG did not partake in these rituals. How could she have contracted Kuru disease?

Credit: Licensed illustration of prions © Oscar Collica | Dreamstime.com, ID 176711652. On left side is the normal form (PrP^{C}) with more alpha helices; on the right side is the infectious form (PrP^{Sc}) with more beta sheets.

2. Based on your knowledge of Kuru disease, what symptoms of Kuru are similar to those of Parkinson's disease leading to the patient's initial diagnosis? What symptoms could the hospital have looked for to distinguish the two?

3. Which medical test could the hospital have conducted to confirm that the disease was Kuru? Why would this test rule out Parkinson's disease?

4. If the ordered blood panel had included a test to determine the presence of prions, would this have changed the initial diagnosis? Why or why not?

5. Given that the motor control medications for Parkinson's did not work for this patient, what other treatments should she have received to treat the Kuru infection?

6. How could have Patient SG's contraction of Kuru disease been prevented in this scenario? Are there other prevention methods?

7. Why do you think the doctors did not suspect Kuru disease? If you could do this case over again, what key information did Patient SG give that could cause a clinician to suspect a diagnosis of Kuru disease?

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