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Cannibals and Mad Cows

The most mysterious epidemic of the XX century

Explorers traveling at the beginning of the XX century in the southeastern parts of New Guinea were puzzled by a strange disease of the locals. Named in the language of the Fore tribe "Kuru" ('trembling with cold and fever') and by Europeans "laughing sickness" due to the outbursts of laughter in its second phase, this disease puzzled the scientists for a long-long time. Three stages were described in the progression of the always fatal disease and the patient died in three to six months. First, an ambulant stage, with unsteadiness of stance, gait, voice, hands, and eyes; deterioration of speech; shivering; loss of coordination in lower extremities and dysarthria (slurring of speech). In the sedentary stage, the patient can no longer walk without support, more severe tremors and loss of coordination of the muscles, shock-like muscle jerks, emotional lability, outbursts of laughter, depression and mental slowing. It is important to note that muscle degeneration has not occurred in this stage, and tendon reflexes are usually still normal. In the last stage, marked by inability to sit up without support; more severe loss of coordination, tremor and dysarthria installs; urinary and fecal incontinence; difficulty swallowing; and deep ulcerations emerge due to brain. By the 50's, kuru was rampant among the Fore tribe, 8,000 individuals community. 1,100 died between 1957 and 1968, mostly women. In fact, eight times more women than men contracted the disease. It later affected small children and the elderly at a high rate as well. Two were the factors that misled the scientists: what provoked the terrible disease and how was the disease transmitted? They first thought it was a genetic condition, as it had a tendency to occur among family members, but kuru was too common and too fatal and such a lethal genetic disorder would drastically reduce the fitness of a population and soon vanish off the gene pool. They injected on chimpanzees brain material from a victim and hypothesized that the agent was a slow virus, as the apes developed kuru after a long incubation period. Thus, at least it was a transmissible infection. Only in 1982 did scientists finally find prions and could explain kuru. Prions are infectious proteins that provoke diseases 100 % fatal. They may be infectious, inherited or spontaneous and the most known prion diseases are mad cow disease (or bovine spongiform encephalopathy) found in cattle and transmissible to humans and Creutzfeldt-Jacob disease, found in humans and very similar in symptoms to kuru, but which develops during at least 30 years. Prions usually attack the brain provoking neurological degeneration (the diseases are called spongiform encephalopathies because the brains usually turn spongy, riddled with holes) and many suspect they are simplified viruses that passed off the nuclear acid phase (DNA or RNA). The researchers were still puzzled by how the prions were transmitted till they discovered the gruesome secret that the tribe was hiding away from the authorities: mortuary cannibalism. When an individual died, the maternal kin were responsible for the dismemberment of the body. The women would remove the arms and feet, strip the limbs of flesh, remove the brains and open the chest in order to get to internal organs. Kuru victims were especially regarded as food, because the layer of fat on victims resembled pork, the most appreciated food item in New Guinea. Women fed morsels, such as human brains and various parts of organs, to their children and the elderly. Kuru was transmitted through participation in such cannibalism or through wounds when removing infectious tissue from the body. The gender disproportion was later traced to the distribution of the corpse's remains between the sexes. The males ate the "good" parts of the body, usually the muscles and fatty organs. The females and children got the "bad" parts, including the brain and the other less desirable parts. This way, the women and children directly ingested the prion, leading to a much higher occurrence rate of Kuru among women. The disease

disappeared with the termination of cannibalism in New Guinea.