Scrapie and kuru.

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Abstract: The suggestion that scrapie, an endemic disease of sheep, might be a myopathy, is rejected by the author, who writes to point out certain similarities between this disease and kuru, a neurodegenerative disease endemic in the Fore tribe of Guinea [this Bulletin, 1959, v. 56, 1283]. Both are endemic in well-defined populations, since scrapie occurs only in certain flocks, and kuru is confined to a single ethnic group, the Fore tribe. Each may develop some months after separation of the individual from his native stock, and each may appear in the progeny of union of affected with previously unaffected stock. In neither case has any recognized bacterial or viral agent been isolated, toxic and nutritional features...
recognizable in either and both appear to be determined genetically. In each course is relentlessly progressive, remission or recovery being very rare. Each are broadly characterized by tremors, ataxia and defects in behaviour. There are certain striking similarities in the histopathology of the central nervous system in each disease. In scrapie, a characteristic finding is the presence of single "soap-bubble" vacuoles in the cytoplasm of affected nerve cells, and similar changes have been observed in kuru. Certain non-specific neuronal degenerative changes are also intense astrocytic activity with minimal myelin degeneration. Inflammation reactions are also unusually meagre for so extensive an "encephalitis", while the cerebrospinal fluid shows no constant or significant abnormalities in either disease.

An important discovery has been that scrapie can be transmitted to previously uninfected stock by the intracerebral or subcutaneous inoculation of brain homogenate from affected sheep, though the manifestations of the disease may not appear for 30.5 to 30 months thereafter. The nature of the transmissible pathogenic factor is so far undiscovered. The author suggests that experiments should be made to transmit kuru from man to a laboratory primate. [Since such an experiment might open the door to further investigation into the aetiology of kuru, which at present has reached an impasse, this seems a most valuable suggestion.]

J. H. Walters.
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