Prions and neurodegenerative disorders.

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Abstract

Only after a protracted and exhausting battle could it have been accepted that a protein might function as an infectious pathogen and lead to central nervous system degeneration. In addition to explaining how a disease can be both infectious and genetic, the concept of prions has also disclosed previously unidentified types of neurological illnesses. The most prevalent neurodegenerative illness is Alzheimer's disease. People in the US suffer from Parkinson's disease and Alzheimer's disease, respectively. Amyotrophic Lateral Sclerosis (ALS), frontoTemporal Dementia (FTD), prion illnesses (including Huntington's disease), and spinocerebellar ataxias are far less frequent. Atypical neuronal protein processing underlies nearly all neurodegenerative diseases. Protein misfolding, altered post-translational modification of freshly generated proteins, incorrect proteolytic cleavage, abnormal gene splicing, improper expression, or decreased clearance of protein degradation can all be components of the aberrant mechanism.

Keywords: Amyotrophic Lateral Sclerosis (ALS), Prions, Frontotemporal dementia, Amyotrophic lateral sclerosis.

Introduction

Infectious proteins are called prions. When prions infect mammals, they attract healthy cellular prion protein (PrPC) and encourage its transformation into the disease-causing (scrapie) variant (PrPSc). The fact that PrPSc is encoded by a chromosomal gene sets prions apart from viruses. Limited proteolysis of PrPSc results in the formation of the smaller, protease-resistant molecule known as PrP, which polymerizes into amyloid. Although PrPC and PrPSc's polypeptide chains are identical in composition, they differ in how they are folded into three dimensions. While PrPSc has a greater proportion of flattened strands of amino acids than PrPC does, PrPC is rich in -helixes, which are spiral-like structures of amino acids. PrPSc may only contain two -helixes and more -strands, in contrast to PrPC, which has three -helixes and two short -strands, according to a feasible hypothesis [1].

Prions are the only infectious pathogens that lack nucleic acid that are known to exist. All other infectious agents have genomes made of either RNA or DNA, and these genomes control how their offspring are made. Second, prion illnesses can present as sporadic, hereditary, or infectious disorders. There is no other category of diseases with a single source that exhibits such a broad range of clinical symptoms. Third, the build-up of PrPSc, which has a significantly different structure from that of its predecessor, PrPC, leads to prion disorders. Fourth, PrPSc can exist in number different conformations, each of which appears to be linked to a particular illness. A wide range of clinical symptoms are present in prion illnesses, including dementia, ataxia, sleeplessness, paraplegia, paraesthesia, and aberrant behaviour [2].

Prions have been classified according to the speed at which they damage the central nervous system and the pattern of neuronal vacuolation. These strains have also been identified using patterns of PrPSc deposition. There is growing evidence that the PrPSc protein's shape encodes the variety of prions. The tertiary and quaternary structures of PrPSc carry strain-specific information, according to studies showing that mice harbouring chimeric human-mouse PrP transgenes can transmit deadly familial insomnia and familial Creutzfeldt-Jakob disease. Somatic mutations have the potential to create sporadic prion disorders, which could then progress similarly to prion diseases brought on by germ-line mutations [3].

Most cases of frontotemporal dementia, Parkinson's disease, amyotrophic lateral sclerosis, and Alzheimer's disease are sporadic, similar to cases of the prion illnesses; although age is the key risk factor for all of these sporadic diseases, it is still unclear what causes neurodegeneration to begin. The initial production of PrPSc in the prion diseases results in an exponential growth of the protein, which is easily transmissible to another host. The circumstances that result in the synthesis of abnormally processed proteins and the mechanisms that maintain their accumulation in the other neurodegenerative illnesses are unknown. It's critical to note that unlike the prion illnesses, frontotemporal dementia, Parkinson's disease, amyotrophic lateral sclerosis, and Alzheimer's disease are not contagious and have not laboratory animals with the transmission [4]. They are:

Alzheimer's disease: Neurofibrillary tangles and A-amyloid plaques can develop in both inherited and spontaneous variants of Alzheimer's disease. Familial Alzheimer's disease

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is inherited in an autosomal dominant manner, just like familial prion illnesses.

Pick's disease and frontotemporal dementia: Pick's disease and inherited forms of frontotemporal dementia are caused by mutations in the tau gene, which produces the protein tau, a microtubule-related protein. Like Alzheimer's disease, frontotemporal dementia is sporadic in around 90% of cases and familial in the remaining 5%.

Parkinson's disease: The majority of Parkinson's disease cases are sporadic; however central nervous system protein deposits are a feature of both sporadic and familial forms of the disease. Patients with familial Parkinson's disease have been shown to have mutations in the syncline gene [5].

Conclusion

There are no effective treatments for neurodegenerative disorders other than levodopa, which reduces Parkinson's disease symptoms but does not stop the underlying degeneration. Very few attempts to stop or stop protein misprocessing have been effective in the past. Creating novel medications that target particular parts of the central nervous system will be difficult. Multiple drugs have been developed as a result of dominant negative suppression of prion formation in structure-based drug discovery. However, it is still challenging to switch from polypeptide scaffolds to tiny heterocyclic structures without losing biologic function. It is unknown if this strategy for avoiding the abnormal processing of proteins will result in the creation of fresh medications for the neurodegenerative diseases Parkinson's, Alzheimer's, and other. Levodopa replacement therapy has been effective because Parkinson's disease neurodegeneration is mostly contained to the substantia nigra, especially early in the disease process. Eventually, however, the disease becomes resistant to levodopa in many individuals. Similar methods to treating Alzheimer's disease have been unsatisfactory, primarily due to how pervasive the disease process is. The broad neuropathological alterations in prion disorders, frontotemporal dementia, and amyotrophic lateral sclerosis also make it unlikely that replacement therapy will be effective.

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