Review Article

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Prions and Oral Cavity: The Oblivious Single Handed Protein Machinery.

Abstract:

Background: The world today is facing an unprecedented situation with the spread of novel coronavirus or covid-19. Covid era has brought the whole world to its knees. Oral health care professionals are at the risk of acquiring numerous diseases which amounts to occupational hazards. Now is the time to widen our horizons and look at different spectrum of diseases, prions being one of them. The short clinician course and long incubation period poses many challenges to this group of neurodegenerative diseases.

Aim: The aim of this review is to highlight uncharted aspects of prions and oral health.

Review details: It sheds light on the spectrum of prion diseases. Since there is no known cure of this dreaded disease and is ineffective with normal sterilization and disinfection procedures, prevention is the only way ahead. This review highlights the colliding world of prions and human body. This also keeps in mind the precautions and guidelines which will help health care professionals in combating this non curable fatal disease. CONCLUSION: Ensuing endeavors should be undertaken to help find plausible detection techniques, treatment options (if any), precaution and guidelines to be kept in mind to fight off this protein entity.

Keywords- prions, health care professionals

Introduction:

During the course of human history, outbreaks of lethal diseases have savaged entire populations, cities and countries. Whether it's the prehistoric times, the historic times or the recent times spread of these disease from countries to countries have crippled the entire human race. Role of humans in constant wiggling with the ecosystem of animals and the constant fluxing between microbes, animals and humans have created new harbors for harboring viruses and in its spread. Henceforth, zoonotic origin diseases are on the rise.

First reported case of scrapie was reported in flock of Scottish sheep in 1939.[1] It was spread after a vaccine was injected to these sheep from ill prepared infected sheep brain extract.[2]

The kuru epidemic in Papua New Guinea was caused by cannibalistic behaviours of the native population by sporadic Creutzfeldt-Jacob disease (CJD).[3] Bovine spongiform encephalopathy (BSE) in 1980's was fuelled by feeding of prion contaminated meat meal to cattle.[4] A case in humans was discovered by Stanley Prusiner who won the nobel prize in medicine in 1997. He defined prions as modified isoforms of infectious transmissible particles that lacked nucleic acid.[5] What led to the conclusion of it being 'only protein extract' was that it is resistant to UV radiation unlike the nucleic acids.[6] Basically, transmissible spongiform encephalopathies (TSE) also known as prions are a group of neurodegenerative diseases found in humans and animals. Names vary according to the host. Its called BSE in cattle,

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scrapie in sheep and goats, chronic wasting disease in (CWD) in deer and CJD in humans.[7]

Transmission:

The transmissibility of these distinct disorders is very interesting. They are not contagious but are transmissible through parental and oral route. While scrapie and CWD can be transmitted within animals, BSE and CJD appear to have limited /no transmission between individuals.[8] Infected placenta has also been suggested.[9] Animal study has shown this disease spreads by dental route.[10]

Molecular Structure of Prions:

PrP is a host protein found predominantly on outer surface of neurons attached by glycosylphosphatidylinositol (GPI) anchor. According to the updated version of protein hypothesis, prion is a conformational isoform (PrPc) of normal host protein PrP.[11] In prion disease largely protease resistant aggregated form of PrP i.e. PrPsc also accumulates in brain.[12] PrPc and PrPsc are different only in their conformational structure not in their primary structure.[13] The beta sheet content of PrPc was low as compared to PrPsc.[14] The two states are separated by activation energy barrier only.[15]

Routes of propagation of prions[12]:

After being ingested orally, prions may penetrate the mucosa of intestine (M cells) and reach Peyer's patches and enteric nervous system. They may also replicate in lymph nodes and spleen. From the lymphoreticular system, they travel to the peripheral nervous system and reach brain (via the vagus nerve or spinal cord).

Studies On Human Oral Tissues

Study by Guiroy et al. have noted presence of PrPsc around nerve root and axons of trigeminal ganglion and trigeminal nerve which could lead to possible extension into oral and nasal cavities. This could serve as a route of transmission via the oral route.[16]

Another study which evaluated post mortem cases of vCJD showed that majority of cases which were positive for PrPsc had ample amount of PrPsc in tonsils and trigeminal ganglion. Immuno-histological techniques employed for this study include paraffin-embedded tissue blot and western blot. These tests indicated that level of PrPsc is at level of less than 1% that found in brain tisssues.[17]

However, a study by Blankuet-Grossard et al. couldn't find any protease-resistant PrPsc in pulp tissue from eight patients with sporadic CJD.[18]

Oral Manifestations:

There are rarely any manifestations. Some laryngeal manifestations include dysphagia and improper speech articulation. They develop due to pseudobulbar dystrophy.[19] Some other vague manifestations have been reported in literature like paresthesia, loss of taste and smell.[20,21]

Management of TSE Patients in Dental Health Care Settings:

Identification of patients with different forms of prion diseases – inherited and acquired (Kuru, iatrogenic CJD, variant CJD, sporadic CJD), knowledge about endemic areas, identification of patients who had travelled to such areas and appropriate history taking.

Below are the many possibilities of transfer of prion infection in dental health care settings:

1. Accidental trauma of lingual tonsils of TSE patient during any dental procedures, subsequent instruments getting infected.
2. Any inadvertent contact with dental pulp tissue.
3. Theoretical possibility of shedding prion via skin as demonstrated in rodents by Thomzig et al

Prions are resistant to most clinical disinfection and sterilization procedures and can only be completely destroyed

by incineration. Care should be taken with regard to endodontic procedures as risk of transmission is higher due to reuse of endodontic instruments without the availability of proper decontamination procedure of prions. It is recommended to use disposable instruments wherever possible and send those instruments for incineration which are difficult to clean.

Conclusion:

Physical and psychological management of such patients deserve special attention. Care should be taken for proper identification of such patients as already mentioned. Dental professionals should always keep themselves updated about the newer variants of biological pathogens in interest of themselves, their patients as well as society at large.

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