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PRIONS IN DENTISTRY- A REVIEW

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Abstract

Prion diseases are immedicable neurodegenerative conditions affecting both animals and humans. The disease was first discovered by Stanley B. Prusiner and he defined Prions as infectious, transmissible proteinaceous particles that lack nucleic acid and are composed exclusively of a modified isoform of the non-infectious cellular prion protein (PrPC). The dentist must bear in mind of this disease because the Prions resist the standard sterilization procedures in normal clinical use and is barely completely eliminated by incineration. This article reviews current knowledge of the presence of prion within the mouth, risk of transmission and discusses infection control protocol for the patients with prion disease.

Keywords: Prions, Creutzfeldt–Jakob disease (CJD), Gerstmann– Straussler–Scheinker disease, Prion Protein.

INTRODUCTION

It has been almost four decades since the discovery of the proteinaceous infectious particle aptly coined as 'Prions' by its discoverer Stanley B. Prusiner. Prions have been known to be associated with multiple diseases. Synonymously, prion diseases are also called as Transmissible spongiform encephalopathies. Prusiner defined prions diseases as infectious, transmissible proteinaceous particles that lack nucleic acid and are composed exclusively of a modified isoform of the non-infectious cellular prion protein (PrPc). The sporadic type of Creutzfeldt-Jakob disease (CJD) is the commonest human prion disease; the mean age of affected is 68 years, the fatality rate is 85% within 1 the average death rate is 1 per million persons. Variant CJD (vCJD) affects people (mean age 26 years) with a history of previous extended periods of residence in certain countries, mainly year, and in the United Kingdom.[1] Transmissible spongiform encephalopathies (TSEs) are malefic neurodegenerative diseases that are rapidly progressive and always deadly, with no approved cure, and their definite diagnosis can only be obtained at post mortem autopsy. Prion protein, resists routine sterilization methods especially when infected tissue becomes dried

onto glass or metal surfaces, which poses a big risk to the patients especially in a dental operatory where instruments are routinely infected and sterilised through common methods. Because there's a real risk of transmission of prion disease from dental instruments (although it's extremely low, especially in North America), as a general rule, appropriate family and medical record (including the danger for prion diseases) should be obtained from all patients, before all dental procedures.[2] Globally annual incidence of CJD was reported as 0.3 to 1.1 per million population. In India, Department of Neuropathology, National Institute of psychological Health and Neurosciences, Bangalore reported an incidence of 0.085 per million.[3] Although prion diseases are rare, the differential diagnosis includes common conditions such as Alzheimer's disease and vascular dementia.[4] This article aims to give a brief overview of risk of transmission and infection curb methods of prion diseases for dentists using data obtained from PubMed search engine.

ETIOPATHOGENESIS

Normally, the Cellular Prion proteins (denoted by PrPC) are present on the surface of cells, particularly on the neuron surface and function during synapses. However, Prions are not similar to PrPC, as they are misfolded version of the normal cellular prion proteins denoted as PrPSc (Scrapie form). The infectious agents for prion disease are composed of a 35-kD brain sialoglycoprotein called PrPSc that's essential for the transmission and pathogenesis of several neurodegenerative diseases. PrPSc propagate itself in the host by stimulating the conversion to PrPSc, resulting in its accumulation.

Any protein in the physiologic system undergoes folding to achieve stable tertiary structures. If there is any deviation in this folding i.e., if there are errors during the folding mechanism then the proteins get misshaped. These misshaped proteins have a correction mechanism called as Chaperone catalysis by which they are again converted to normal folded proteins. Now, if these misshaped proteins are not corrected by the chaperone molecules, then they are sent for degradation by proteolysis. When it comes to misshaped prion proteins, they have resistance to proteolysis and therefore remain undegraded in the body which causes an inadvertent accumulation of PrPSc which may also convert normal PrPC to abnormal misfolded PrPSc proteins.

Accumulation of PrPSc may result either from exposure to infectious prions iatrogenically or through ingestion, or due to mutations in the PrP gene. Sporadic CJD has an unknown cause; So far, no apparent infections or mutations of the PrP gene are found in association with such cases, although the brain in these patients also accumulates PrPSc.[5]

ORAL MANIFESTATIONS

Oral manifestations are rare and seem to occur only in people with CJD (Creutzfeldt-Jakob disease); there are no oral mucosal or gingival manifestations of prion disease. Prions can be detected in the oral tissues—usually the gingivae and dental pulp—of animals experimentally infected with prions.[6] Dysphagia (difficulty in swallowing) and dysarthria (poor articulation of speech), paraesthesia (tingling, pricking or numbness), orofacial dysesthesia (abnormal sensations in the absence of stimulation) and in one case loss of taste and smell have been reported in the literature.[7]

INFECTION CONTROL IN DENTISTRY

TSEs do not spread by contact between persons, however during invasive surgeries and medical interventions there is a high risk of transmission. The tissues in the oral cavity such as the gingiva and saliva do not pose any detectable infectivity but certain procedures involving the eye, brain or the CSF has high risk of prion transmission. Laboratory studies indicate that standard decontamination and

sterilization procedures could also be insufficient to completely remove infectivity from prion-contaminated instruments. The instruments may spread the prion disease to others. Much caution therefore should be taken within the absence of strong evidence against the presence of a prion disease in a neurosurgical patient.[8] The participants of WHO Consultation on Infection Control Guidelines for TSEs suggested that single-use items and equipment like disposable needles and anesthetic cartridges represented the safest method for minimizing the exposure of prions. The neurovascular procedures and instruments used in these procedures should be incinerated or decontaminated in combination with chemical and autoclave techniques.[9][7] In patients with suspicion of CJD, all the instruments must be stored separately in a very rigid container labelled with data of the patient, treatment provided and details of the attending clinician until a specific diagnosis is ruled out. The instruments are incinerated if the findings are established or disinfected by conventional methods like autoclaving if conclusion is precluded. [5]

DISCUSSION

As of late, there has been an expansion in logical and public mindfulness about prion infection. There appears to be a very low likely danger of CJD transmission in routine dental practice with more risk of transmission only in certain cases of oral and maxillofacial surgery. Since there is no scientific evidence of transmission by direct contact between people or via air or saliva, the only special requirement in the dental office is for measures to be taken to inactivate prion on instruments which should be done following the recommendations that has been explained before. The quick advancement of the disease and the physical and mental reliance of these patients may mean that they require special dental treatments. The exact nature and structure of prion and the danger of transmission of prion disease through dental therapy focuses to the significance of keeping up ideal principles of infection control and decontamination for infectious agents, including prions. So, the dental experts should have up-to-date knowledge about transmission, diagnosis, contamination control and decontamination procedures regarding prion diseases.

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