Editorial

Standardized surveillance of prion diseases in resource-poor settings is crucial for individual patient-care as well as for decision-making of healthcare authorities

In their present study Mahale and colleagues report a series of eight Creutzfeldt-Jakob disease (CJD) patients collected over 3 years in a tertiary care hospital in Bangalore.[1] They thereby present the fourth published CJD case series from India raising the total number of identified patients to just over 100.[2-4] Under the assumption that incidence rates are comparable to those in the rest of the world (about 1:1,000,000), about 1000 cases of sporadic CJD per year would be expected in India (given a total population size of 1 billion). The evidence for stable incidence rates irrespective of the geographical setting is, however, primarily based on data from genetically and environmentally homogeneous populations (Europe, North America, and Australia). From an epidemiological point of view it would thus be very interesting to investigate if incidence rates in India differ from those in other countries (e.g. for genetic or environmental reasons). Unfortunately, this cannot be assessed in the present setting as there is no standardized nationwide surveillance for human prion diseases in India. The observed discrepancy between 100 reported cases within the past three decades and 1000 expected cases per year has to be attributed to surveillance bias rather than to unexpectedly low incidence rates in the Indian population. Intensity of surveillance programs has been shown to directly predict reported CJD incidence rates in those countries which have nationwide surveillance strategies.[5] In countries, which haven't (like India), incidence rates close to 0 are commonly reported. In these settings observed incidence rates are not useful for describing the burden of disease. This is especially difficult if surveillance patterns change over time resulting in increasing incidence rates; true outbreaks are hard to spot in such a complex temporo-spatial environment. Therefore, standardized surveillance programs need to be established in order to empower healthcare authorities for making evidence-based decisions.

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Standards for the surveillance of human prion diseases have been set by the implementation of prion surveillance units in many Western countries following the epidemic of variant Creutzfeldt-Jakob (vCJD) disease and the Bovine Spongiform Encephalopathy (BSE) crisis in the 1980ies and early 1990ies. Health authorities recognized for the first time the potential pandemic threat of human prion diseases and implemented surveillance units for active case-finding strategies in order to prevent the next outbreak. Active surveillance (including home visits at patients with suspected CJD and active follow-up of those referred for 14-3-3 testing) is crucial for diseases with low incidence rates as expert knowledge is necessary for a proper diagnostic work-up and a correct classification.

However, these newly established surveillance units were not only able to increase the knowledge of public health authorities about the incidence of prion-like diseases in the population, but did also improve individual patient-care. They increased physicians' awareness for the diseases, supported them in the diagnostic work-up with standardized procedures and expert knowledge, supported the affected families with their experiences in patient-care and developed new diagnostic tests and standardized criteria for a better diagnostic process. As all of the units were faced with low numbers of patients, they started working closely together and supported each other, e.g. in the validation of tests or by performing ring trials and thus improved patient-care once more. These high standards are the aim for all countries which start contributing to worldwide prion surveillance. Although there are still many aspects missing (e.g. a certified laboratory which can perform standardized 14-3-3 testing and takes place

in multinational ring trials), India is on a good way to implement these high standards.

Thus, the establishment of a standardized surveillance program for prion diseases in India is crucial for detecting changes in prion disease incidence rates over time, for investigating differences between prion disease incidence rates in India and those in other countries and for improving individual patient-care for all individuals who face this devastating disease.

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