Chapter 14
Lewy Body Disease:
Point Towards Progressive Dementia

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ABSTRACT

Fritz Heinrich Lewy described the intracytoplasmic inclusions found in the neurons for the very first time. In 1919 these inclusions were termed as "LBs" by Tretiakoff. LBs were found in the brain of the patients suffering from Lewy body disease (LBD). LBD is characterized by the presence of Parkinsonian symptoms in the earlier stages and dementia in the later stages of the disease. LBs were classified on the basis of the region of the brain in which they are distributed and so is the case of the LBD means the type of the LBD depends on the anatomical areas of the brain involved. LBD is not a single disorder. It is a spectrum of disorders. This chapter addresses the entire profile of LBs, types, composition, formation, and various LB pathologies as well as diagnostic criteria and pharmacotherapy.

INTRODUCTION

Lewy bodies (LBs) are the eosinophilic intracytoplasmic inclusions found in the brain of the patients of Parkinson's disease (PD) and were first described by Fritz Heinrich Lewy. These inclusions were later called "LBs" by Tretiakoff in 1919 (Wakabayashi et al, 2007). The presence of LBs in the brain of PD patients suggesting the possible involvement of LBs in the process of neurodegeneration, but the recent reports suggested that the LBs formation is a response evoked by the body to scavenge or degrade the neurotoxic substances that is responsible for neuronal degeneration (Sathiyamoorthy et al., 2014). LBs occur in context of various distinct pathologies known as Lewy body disease (LBD) characterized by the presence of the parkinsonian symptoms in earlier stages and dementia in the later stages of disease. Various symptoms of LBD include parkinsonism, hallucination, cognitive impairment, fluctuations in mental status, and dementia. Different types of LBD shares some common characteristics however the marked symptomatic difference found in these pathologies is mainly due to the difference in anatomi-

DOI: 10.4018/978-1-5225-5282-6.ch014

cal region of the brain involved (in which the LBs formation occurs) (Kosaka, 2014). Further each of these pathologies is characterized by the presence of LBs, therefore the symptoms appears very similar and sometimes overlaps making the exact pathology difficult to diagnose. Thus the exact pathology so involved sometimes remains undiagnosed and often misdiagnosed (Galvin & Balasubramaniam, 2013) which often becomes troublesome to the patients and caregivers (Galvin et al, 2010). Therefore very precise diagnostic criteria's is required to avoid the false diagnosis and the wrong prescription that may prove fatal later. The difference in diagnosis largely depends on the order of symptomatic presentation such that if only the movement deficits are present then the person is said to be suffering from PD, the development of dementia one year after the onset of the PD is referred as dementia associated with PD (PDD) and if the dementia develops before the onset of the PD or within one year of the PD then it is referred as dementia with LBs (DLB) (Galvin & Balasubramaniam, 2013). In the present chapter authors demonstrate LBD with their similar and differentiating characteristics and provides several recommendations that will prove beneficial in the diagnosis and the pharmacotherapeutics of such pathologies.

BACKGROUND

LBs are the intracytoplasmic inclusions found in the neurons of the patients suffering from LBD, however the exact type of the LBD which a patients exactly suffers from depends upon the region of brain in which the LBs were found predominantly (Kalra et al, 1996). Brainstem nuclei involvement is almost universal in the LBD while the involvement of limbic region and neocortex suggested the progression of disease (Braak et al, 2003). Further it is suggested that the brainstem is involved in the early stages of LBs pathologies whereas the different region of brain also involved when the disease progresses or in the later stages of the disease (Adler et al, 2010). LBs pathologies or the LBD represent a family of the disease that involved three distinct pathologies including PD, PDD and DLB. Before going in to the depth involved in the pathogenesis of these pathologies it remains of interest how LBs occurs and what is the reason of LBs formation. Recent reports suggested that the dopaminergic neurons isolated from the healthy individuals when implanted in the brain of the PD patients form LBs (Kordower et al, 2008; Li et al, 2008) suggesting LBs formations occurs to counteract the pathological insult. Thus LBs formation represents an epiphenomenon, or the scars of neurodegeneration (Popescu et al, 2004). In the present work authors demonstrate the different types of LBs, their formation and composition and the occurrence of LBs in various LBD.

CLASSIFICATION OF LEWY BODIES

LBs can be classified on the basis of their distribution in the different regions of brain.

Type 1 Lewy Bodies

Type 1 LBs are also known as brainstem LBs because of the predominant involvement of the brainstem nuclei (Braak & Braak, 2000; Dickson et al, 1987; 1996; Kosaka et al, 1984). Type 1 LBs are spherical in shape containing eosinophilic cores, lamellar bands and pale halos (Campbell et al, 2001).