Volume-4, Issue-4, April-2015 • ISSN No 2277 - 8160

Research Paper MEDICAL SCIENCE

Visuo-Perceptive and Visuo-Constructional Deficits in Lewy Body Disease

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ABSTRACT

Introduction and Background: Dementia is a chronic neuro-degenerative disorder characterized by progressive cognitive loss. Alzheimer's disease and the Lewy Body disease are the two most common causes ofage-related degenerative dementia. Visuo-cognitive skills are a combination of very different cognitive functions being performed by the visual system. These skills are impaired in both AD and DLB. The aim of this review is to evaluate various studies for these visuo-cognitive skills. Methodology: An exhaustive internet search of all relevant medical databases was carried out using a series of key-word applications, including The Cochrane Library, MEDLINE, PSYCHINFO, EMBASE, CINAHL, AMED, SportDiscus, Science Citation Index, Index to Theses, ZETOC, PEDro, and OT Seeker, and OT search. We reviewed all the articles till March 2013 with key words of: visual skills, visual cognition, Dementia, Alzheimers disease, Dementia of Lewy Body disease. Results: Although most studies have used different tests for studying these abilities, in general, these tests evaluated the individual's ability of 1) Visual recognition 2) Visual Discrimination 3) Visual attention and 4) Visuo-perceptive integration. Performance on various tests has been evaluated for assessing these skills. Most studies assessing such skills show that these skills are impaired in DLB as compared to AD. Conclusion: Visuo-cognitive skills are impaired in DLB. These impairments have evident neuropathological correlations, but the direct neurobiological etiology is difficult to establish.

KEYWORDS : LEWY BODY DISEASE; VISUOPERCEPTIVE ABNORMALITY; VISUOCOGNITIVE ABNORMALITY

INTRODUCTION

Dementia is an umbrella term used for a group of chronic neuro-degenerative disorders with a common characteristic of progressive cognitive loss. Alzheimer's disease and the Lewy Body disease are the two most common causes of degenerative dementia. Visuo-cognitive skills are a combination of diverse cognitive functions involving the visual system. Presence of these skills is pivotal for day to day activities. These skills are much more important from the perspective of a clinician facing the challenges of treatment of neuro-degenerative dementia patients in view of the fact that impairment of visuo-cognitive skills is a common component of these Dementias (Tiraboschi et al., 2006). In this review, we will focus on the impairments of these skills in LBD. A clinical neurologist dealing with LBD not only requires this knowledge to properly guide the patient through treatment, but also to diagnose this disorder in the first place. From this clinical stance, there are three important reasons regarding why these skills need to be properly addressed. The first and perhaps the most important from the patient's perspective is the imperative requirement of visual skills in performing day to day activities. This imperativeness becomes evident in the Lewy Body Disease patients where difficulties in VC skills is known to create several troublesome situations. In other forms of dementia also, there are impairments in these skills. For example, in advanced cases of AD, there is a loss of navigational skills which constitute an important part of day to day activities, especially in out-of-the-home circumstances. Second important reason is the diagnostic implication of such visual skills in identifying Dementia with Lewy Bodies (DLB). This diagnostic implication is often a priority, given reports of severe neuroleptic sensitivity and a preferential response to cholinesterase inhibitors in these patients. There have been suggestions that constructional apraxia is prevalent in DLB, and may provide a sensitive marker of the disease (Cormack et al., 2004). In fact, visuoconstructional tasks, in combination with other tests, can differentiate DLB from normal aging and from AD with sensitivity in excess of 80% and specificity in excess of 90% (Ferman et al., 2006). Because impairment of visual abilities in DLB is a consistent finding, it has also been suggested that visuospatial deficits may actually be a particularly salient biological marker of DLB (Tiraboschi et al., 2006). Finally, the third clinically important reason is the implication of these skills in evaluating the severity and prognosis of DLB. It has been suggested that visuo-cognitive skill deficits are related to presence of other symptoms of the disease, especially the Visual hallucinations. Studies have shown that perceptual deficits evident in DLB may be responsible for some misperceptions (i.e., illusions) and delusional misidentification (i.e., not recognizing family, reduplicative paramnesia). Interestingly, DLB patients with visual hallucinations tend to perform more poorly on visual tasks than the subset of patients not having VHs (Ferman et al., 2006; Calderon et al., 2001). Identification of visual spatial impairment is important not only for designating individuals whose clinical syndrome is impacted more by Lewy body formation than AD pathology, but also for predicting which patients with DLB will have a more malignant disease course (Hamilton et al., 2008). Throughout this review, we will evaluate these impairments in the background of these three dimensions.

DEFICITS IN VISUO-PERCEPTIVE SKILLS

Behind our unique perceptual abilities lie several complex and dynamic interactions of a number of basic visuo-perceptive skills. Different studies have defined and categorized these basic perceptual skills in different ways so that a unified picture encompassing all of them remains to be developed. This situation seems to be because the studies have applied different tests for the assessment purposes of cognitive functions. Overall, these test domains can be divided into the basic abilities of 1) Visual recognition 2) Visual Discrimination 3) Visual attention and 4) Visuo-perceptive integration (Calderon et al., 2001; Mori et al., 2000; Mosimann et al., 2004). However, these tests have often tap more than one of the above-mentioned domains. Furthermore, these studies have used different terminologies to denote these visuocognitive abilities. Thus, for the purpose of this review, we will evaluate one study at a time in order to avoid any confusion arising from terminologies and concepts.

Calderon et al. (2001) used the screening test (figure-ground discrimination) from the Visual Object and Space Perception (VOSP) to differentiate the elementary visual identification performances of patients with DLB, AD, and normal controls. They found that all the three groups showed similarly performances for thereby implying that the patients with AD as well as DLB were able to understand the task thus retaining the basic levels of visuoperceptual abilities. However, on three of the subtests of the same VOSP test namely (a) fragmented letters (b) object decision and (c) cube analysis, there were statistically significant differences between the groups as revealed by the post hoc analyses such that there were impairments in the DLB group when compared to both controls as well as the patients with AD. Authors attempted to explain these findings on the basis of the commonly used dichotomy of input processings: Those processing shape properties (ventral system) and those responsible for processing spatial properties (dorsal system) (Goodale & Milner 1992). Their results depicted that DLB patients had more severe impairments on tests evaluating the functions of both ventral stream (Fragmented Letters and Object Decision) and dorsal stream (Cube Analysis) as compared to the AD patients . However, no difference was observed between controls and the AD group. On the silhouette identification test the AD and DLB groups were equally impaired as compared to the controls. This task requires the subject to identify the silhouette profile of the specific objects and animals when viewed from different perspectives. It gives and indication of both the perceptual and semantic

performances of the individual. However, there were other tests applied in the study from the VOSP like the screening test that do not require semantic processes for their completion. The authors have categorized these,as pure tests of perceptual ability. In the study, the patients with DLB failed this test consistently. Similar to these results, Mori et al. (2000) found that both higher-order as well as elementary visual perception (Size discrimination task) are impaired in DLB. Their study showed that DLB patients performed poorly both on the test of elementary visual perception (size discrimination) as well as on the test of complex visuo-perceptual functioning which requires analyzing the 2-dimensional visual stimuli (Form discrimination).

A study focused on assessing Visuo-perceptive skills and for evaluating the effect of psychosis on the same was conducted by Simard et al. (2003). In this study, the Benton Judgment of Line Orientation (BJLO) test was administered to 4 DLB patients with predominant psychosis (DLB-psy) and was compared with 4 DLB subjects with predominant parkinsonian features (DLBPD), as well as with 13 patients with AD. The researchers also applied an analysis of error- types to the results of visual attention errors on the BJLO , as well as to visual-spatial perception errors. The results showed that DLB patients with psychosis committed significantly more errors of visual attention as well as of visuo-spatial perception as compared to the AD patients. However, n total score of the BJLO was not statistically different between the groups. These results have replicated previous findings of visuo-perceptive impairments in DLB and have additionally provided the first evidences for the possibility that not only the the defective visual perception could be a problem in itself but could be a contributing factor for the development of visual hallucinations in such patients. Another study whose findings correlated psychosis with visuo-perceptive deficits was conducted by Mosimann et al. (2004) who tested patients suffering from DLB and PDD with tests for the functions of the theoretical ventral stream (tests of object and form perception) and dorsal streams of (tests of dot position and motion perception) of visual perception. Their results depictedthat both the grops of LBD and PDD had significant impairments in the performanceson tasks requiring visual discrimination, space-motion, and object form perception. They also found that patients with PDD and DLB with visual hallucinations performed significantly worse that the patients without hallucinations. With this background of researches conducted for the visuo-perceptive skill deficits in LBD and AD, their implications in everyday situations can be easily comprehended. Visuo-perceptive skills are primarily required for the identification of persons as well as objects, both of which seem to be impaired in DLB. Visual discrimination on the other hand requires the ability to detect the difference between two similar looking visual stimuli. Thus based on these study results, it can be understood that DLB patients have higher chances of getting lost on the ways outside the home or at times even inside the home especially as compared to AD patients.

To summarize, both elementary and higher order complex perceptual abilities are impaired in LBD as well as in AD. These deficits are more severe in DLB patients with psychosis as compared to those without psychosis, more specifically visual hallucinations. Most studies demonstrate that the impairment is more severe in LBD as compared to AD patients. However, few studies have addressed this issue and the most studies have made assessments on relatively small sample sizes. More studies with better study designs and with larger sample sizes are needed for a better and more authentic results of this domain.

DEFICITS IN VISUAL CONSTRUCTIONAL SKILLS

The issue of diagnostic clarification is specifically important because in later stages of all forms of cortical dementias, the impairment of the constructional abilities starts setting in. Therefore, if detecting impairments in the in early stages can provide any diagnostic clue, it would be valuable from clinical perspective.

Cormack et al. (2004) conducted a cross-sectional study of examining the pentagon drawings of DLB patients, Alzheimers disease (AD) patients, and Parkinsons disease (PD) patients. The performance scores on this task was also correlated with scores on the scales of on the MMSE and CAMCOG . Patients with DLB performed significant worse on pentagon drawing task than the patients with AD or PD, but not patients with PDD. Drawing scores showed statistically significant correlations with MMSE scores for the AD and PDD groups but not in patients with DLB. Further exploration of the cognitive correlates of constructional performance in patients with AD and DLB was performed and the results revealed that patients with AD had a broad possibilities of cognitive basis , but in DLB patients, drawing was associated mostly with the perception as well as the praxis abilities . The next important issue is that of level of brain involvement in the dementia (cortical vs subcortical). The impairment of constructive abilities in LBD is indicated by the poorer performance on the construction subscale performance in patients with Parkinson disease as compared to the patients with AD (Paolo et al., 1995). This finding was replicated other studies as well (Hansen et al., 1990; Sahgal et al., 1995) showing visuospatial and constructional deficits in patients with LBV. Interestingly, one of the most difficult items from the Initiation/Perseveration subscale which was copying the ramparts figure was significantly more impaired in patients with LBV than in patients with pure AD across the full MDRS range. Visuo-spatial praxis is an umbrella term used to describe alll the cognitive abilities utilised in constructional abilities. Several studies have documented impairment in visuospatial praxis in both AD and DLB (Galasko et al., 1996; Salmon et al., 1996; Shimomura, 1998). Most investigators (Walker et al., 1997; Gnanalingam, 1997; Ala et al.,2001; Collerton et al., 2003; Simard et al., 2003; Cormack et al., 2004; Noe et al., 2004), have found a greater visuospatial/constructional (and visual-perceptual) impairment in patients with DLB as compared to other forms of dementia. Copying a diagram requires a coordinated activity of several cognitive domainsAla et al. (2001) found that nearly 59% of DLB patients were definite of producing an unacceptable copy of the pentagon based on the sensitivity analysis. Similarly, in the study by Tiraboschi et al. (2006), visuospatial/constructional functioning was evaluated based on the performance on the MMSE intersecting pentagons and/or DRS construction subscale (DRS-C). Results showed that lThey found that differences in the frequency of flawed MMSE pentagon copying between DLB and Alzheimer's disease patients was statistically significant (30% versus 16%, $p = .01$), the subjects of DLB group also showed a significantly more impairments in the performance on the DRS-C subscale (74% versus 45%, $p = .01$). An erroneous 'vertical lines' reproduction was the most frequent mistake in both groups. Fewer DLB subjects (30%) experienced the difficulty with the MMSE pentagon copy, suggesting that the DRS-C may be more sensitive to visuospatial/constructional impairment early in the course of the disease. In a relatively recent study, Mondon et al (2007) found impaired performances of both DLB and PDD patients on the Rey Osterieth complex figure test (Copy part) and the performance differences were not statistically significant.

Drawing ability is another domain which seems to be particularly impaired in DLB. Similar to the study by Salmon et al (1996), , in the study by Noe et al. (2004), the DLB group performed poorly on the Rosen Drawing Test.

To summarize, DLB patients have severe impairments in visuo-constructional skills. Till this date, this skill has been mainly judged on the basis of some paper-pencil tests like copying and drawing. More sophisticated tests may be required to observe these impairments further closely.

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