When It’s Not Alzheimer’s Disease: Dementia with Lewy Bodies

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KEY POINTS:
- Dementia with Lewy Bodies (DLB) is a neurodegenerative condition, which causes cognitive loss, parkinsonism, visual hallucinations, and REM Behavioural Sleep Disorder
- A trial of cholinesterase inhibitor therapy should be attempted in cases of DLB

With our rapidly aging population, the modern medical practitioner will be confronted increasingly with dementia, given the prevalence of this debilitating, and largely neurodegenerative, class of disorders.

There are five common causes of dementia: Alzheimer’s disease, Lewy Body disease, Frontotemporal degeneration, Parkinson’s disease, and Vascular disease. Despite extensive research at our disposal, differentiating between these causes is rarely an easy task.

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Webinar for Family Physicians

Topic: Hallucinations in the Elderly
Presenter: Lisa McMurray, MD, FRCPC
Royal Ottawa Mental Health Center
Date/Time: Wednesday, January 16, 2013 12 noon to 1pm

Use the button to register

Technical Requirements:
- Visual Support — The presentation will be accessible via an internet connection. This connection can be any web-enabled laptop or desktop computer of your choice.
- Audio Support — Audio support for the presentation will be provided through your telephone via a toll-free line.

You will receive a confirmation email 24-48 hours prior to the session. Thank you, we look forward to your participation!
Here, we deal with differentiating between Alzheimer’s disease (AD) and Lewy body disease, which causes Dementia with Lewy Bodies (DLB). Alzheimer’s disease is known to be the most common of the dementing illnesses, and DLB follows a close second. Importantly, both AD and DLB are criterion-based diagnoses, and are classified as probable or possible at the time of presentation, and definite at autopsy. Many cases of DLB are diagnosed as AD clinically, while postmortem studies show that up to 40% of demented patients have histopathological evidence of DLB.¹

The clinical diagnosis of Alzheimer’s disease requires the presence of progressive memory impairment, and additional impairment in one or more cognitive functions such as apraxia, agnosia, aphasia, or executive function. There must be evidence of decline which has impacted the patient’s social or occupational functioning.² Importantly, systemic (i.e. metabolic, drugs, infection, and structural lesions), as well as psychiatric causes of dementia must be excluded.

Dementia with Lewy bodies is a neurodegenerative disorder of abnormal alpha-synuclein accumulation in the central nervous system. In addition to dementia, it has three core features: spontaneous parkinsonism, recurrent well-formed visual hallucinations, and fluctuating attention and concentration (over minutes, hours, or even days). There are supportive clinical features such as rapid eye movement (REM) sleep behaviour disorder (characterized by repeated flailing of limbs or acting out during dream activity), falls, syncope, autonomic dysfunction, delusions, and depression. DLB may show striking neuroleptic sensitivity, such that treatment with antipsychotic medication requires considerable forethought.⁴

Neuropsychological testing in DLB often shows prominent visuospatial or executive dysfunction, as compared to AD, which shows prominent short-term memory decline. DLB generally follows a more rapid course compared to AD, with more psychiatric pathology, shorter time to nursing home placement, and a shortened survival.

Importantly, cases of DLB may be quite responsive to cholinesterase inhibitor therapy (e.g. Aricept/donepezil, Reminyl/galantamine ER, or Exelon/rivastigmine), so a course of this medication class should be attempted.

Considerable research has been made into enhancing the diagnosis of AD and DLB with biomarkers such as cerebrospinal fluid and PET imaging. Ultimately, the diagnosis remains a clinical one, with cognitive testing, medical work-up, and neuroimaging.

REFERENCES
1. Tarawneh, Rawan, Galvin, James E. Distinguishing Lewy body dementias from Alzheimer’s Disease in Expert Review of Neurotherapeutics. 7.11 (Nov. 2007) p1499

FOR FAMILIES:
Lewy Body info sheet from Alzheimer Canada
CLICK HERE
Mrs. H is a 76 year-old widow who lives independently, is active in her community, and is an important support to her son who has schizophrenia. She has severe bilateral hearing loss, but is otherwise healthy and takes only vitamin D 1000 IU daily. She comes into the ER acutely paranoid, believing people are breaking into her home, hearing multiple people conversing, and hearing classical music playing. She is extremely distressed and agitated.

New onset of hallucinations, and/or psychotic symptoms, should be considered delirium-related until proven otherwise. A thorough physical examination is essential. Basic bloodwork should include CBC, electrolytes, glucose, renal function, LFTs, TSH, calcium, vitamin B12 levels, and urinalysis. Further investigations, such as CXR, ECG, and CT brain may be indicated based on history and physical exam findings. Consider polypharmacy as a possible etiology of delirium, as well as alcohol or benzodiazepine intoxication or withdrawal. Fifty percent of delirium cases have no identifiable cause. Even when you are convinced delirium has been ruled out, the differential remains broad:

- Hallucinations related to dementia (visual more common than auditory)
- Depression with psychotic features (hallucinations uncommon, delusions more likely)
- Mania with psychotic features (delusions more common)
- Primary psychotic disorder
  - Late-onset schizophrenia
  - Delusional disorder
- Secondary to severe sensory impairment
  - Charles Bonnet syndrome (visual hallucinations) / Musical hallucinosis
- Complex-Partial seizures

Treatment depends on the underlying cause. For dementia-related hallucinations, firstly, non-pharmacological interventions should be implemented, tailored to each individual case. Using a behaviour observation scale (like the Dementia Observation System), one can identify patterns of symptom worsening, thus highlighting potential triggers and ways to intervene (www.piecescanada.com).

If such interventions fail to provide optimal reduction in symptoms, and distress remains elevated, then antipsychotic treatment can be considered. Antipsychotic treatment is indicated in cases of primary psychotic disorder, and can be added to antidepressant therapy in mood disorder with psychosis. There is no firm rule when deciding which antipsychotic to use. However, risperidone is a popular choice because its side-effect profile is less anticholinergic, and less sedating.

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HALLUCINATIONS IN AN ELDERLY PATIENT

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At higher doses, extrapyramidal symptoms (EPS) are more likely in risperidone than with other atypical antipsychotics. Dosing should start at 0.25 mg Qhs and titrated by 0.25 mg every 1-2 weeks to a maximum of 1 mg total daily dose. Using BID dosing may help reduce side effects. Watch for increased agitation, which could signal the presence of akathesia. Antipsychotics are not indicated for the treatment of hallucinosis secondary to sensory impairment.

You see Mrs. H a month later. You learn from the discharge summary that she was treated for a UTI with nitrofurantoin. Risperidone 1mg BID was started concurrently. MMSE at discharge was 29/30. She still hears music, though this has been the case for over a decade, and does not distress her. The voices are now fewer and less intense, and only present when she is idle. Paranoia has completely resolved. You notice mild masked facies and rigidity on physical exam. You believe she has musical hallucinosis secondary to severe hearing loss, and auditory hallucinations due to resolving UTI-related delirium. You are concerned about EPS and decide to taper and discontinue risperidone over the next month. Four weeks later, she reports that the voices are worse and she is moderately distressed by these. You conclude she has a late-onset primary psychotic disorder (with a positive family history) and restart risperidone at 0.25 mg qhs. She has a good response without side effects. In addition to monitoring for EPS, the patient’s weight, fasting glucose and lipids should be assessed every 6 months.

REFERENCES


FOR FAMILIES:

Delusions and Hallucinations info sheet from Alzheimer Canada
CLICK HERE

RESOURCES

First Link®, a program that gives patients with dementia and their caregivers and family a direct connection to information and services in their community.
CLICK HERE

A Guide to Scheduling and Billing for Family Physicians (in Ontario)
CLICK HERE