

Dementia with Lewy bodies in Meige Syndrome

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Innov Clin Neurosci. 2012;9(7–8):39–41



FUNDING: No funding was received for the preparation of this article.

FINANCIAL DISCLOSURES: None of the authors have a conflict of interest relevant to the content of this article.

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KEY WORDS: Dementia with Lewy bodies, Meige syndrome, blepharospasm, oromandibular dystonia

ABSTRACT

Meige syndrome is a rare form of segmental dystonia characterized by blepharospasm and oromandibular dystonia. A few case reports of Meige syndrome have been associated with Lewy body pathologies, and the syndrome was also proposed for inclusion in the spectrum of Lewy body disease. We report a case of an elderly gentleman with a history of Meige syndrome for more than 10 years who developed dementia with Lewy bodies. Updated clinical and pathological evidence of linkages between these two conditions is also presented.

INTRODUCTION

Meige syndrome is a form of idiopathic cranial dystonia, which is characterized by oromandibular dystonia and symmetrical blepharospasm.¹ It was first described in French literature in 1910 by Henri Meige.² The disease typically develops between 30 and 70 years of age and is more common in women. It affects facial expression and can cause social and cosmetic disturbances. Uncontrollable bilateral closure of the eyelids may even cause visual impairment. Meige syndrome should not be confused with “Meigs” syndrome, which refers to the triad of ovarian tumor, ascites, and hydrothorax.³

Oral medications³ (anticholinergics, benzodiazepines,

dopamine precursors, dopamine receptor agonists, neuroleptics, and anticonvulsants) and botulinum toxins⁴ are treatment options for Meige syndrome. Unfortunately, the effect of botulinum toxins is only temporary, and the magnitude of improvement obtained through oral medications is at best modest.⁵ More recently, deep brain stimulation has garnered increasing attention as another therapeutic option.^{6,7}

The etiology of Meige syndrome is yet to be established.

Neurophysiological studies have demonstrated abnormal excitability of brainstem reflexes, primary motor cortex, and increased tactile sensory discrimination threshold. Functional imaging also showed altered activity levels in the cerebral cortex and basal ganglia.⁸ Case reports of postmortem cases found Lewy bodies (LB) in the brainstem and basal ganglia of some patients with Meige syndrome. Although the relationship between LB and the development of Meige syndrome is not known, it has been reported that up to 25 percent of brains with Meige syndrome showed LB.⁹ Therefore, it was once proposed to expand the clinical spectrum of LB disease to include Meige syndrome.⁹ Dementia with Lewy bodies (DLB) is a form of dementia characterized by recurrent, vivid, visual hallucinations, fluctuating cognition, and spontaneous parkinsonism or neuroleptic sensitivity.

Pathologically, DLB is one form of LB disease with the presence of cortical and sub-cortical LB.¹⁰ We describe a case of a Chinese elderly gentleman who developed DLB more than 10 years after the diagnosis of Meige syndrome.

CASE REPORT

Mr. N was 67 years old when he presented to neurologists in 1999 with a two-year history of involuntary facial movement and difficulty in food intake and mastication. Physical examination found clonic blepharospasm and orofacial dyskinesia. Investigations, including computed tomography (CT) of the brain, were unremarkable. He was diagnosed with Meige syndrome. The patient was initially fearful about receiving botulinum toxin. He was administered haloperidol 3mg daily for the muscle spasms, with partial effect. Further upward titration of haloperidol dose resulted in extrapyramidal side effects (EPSE). Trihexyphenidyl was added for antiparkinsonism effect. The patient was put on botulinum toxin injections for a short period of time but he refused long-term use. He was maintained on haloperidol 2.5mg three times a day and trihexyphenidyl 1mg three times a day since 2001. He had static right-hand tremor while on this regimen.

In October of 2010, Mr. N was referred for psychiatric assessment presenting with three months of vivid visual hallucinations. Further exploration revealed gradual memory impairment since 2008, with a recent marked deterioration accompanying the visual hallucinations. He had deterioration in activities of daily living and self care. He had worsening right hand tremor and unsteady gait, despite no adjustment of the haloperidol dosage. He sustained a fall fracturing his left humerus in June of 2010. Subsequently, he required nursing-home care. His orientation was poor in the home. He started to have fluctuating, vivid, visual hallucinations in July of 2010, seeing

four people riding in a boat. These hallucinations subsided two days later but recurred in mid-September, leading to referral to our out-patient psychiatric unit.

Upon examination, Mr. N was alert and calm. His speech was scanty and reduced in flow. He was euthymic. He had no active hallucinations or delusions. He was poorly oriented to time, place, and person. He had a mini-mental state examination (MMSE) score of 12/30. Physical examination documented no blepharospasm or oromandibular spasms. Prominent resting tremors in all four limbs and cogwheel rigidity were noted. His gait was shuffling. We gave the patient lorazepam 0.5mg at night when necessary to help his sleep, and offered further follow up.

Later that month (October 2010), the patient was admitted to a medical unit for acute retention of urine after an episode of urinary-tract infection. He was unable to be weaned off the Foley catheter. Our opinion was sought to review his drug regimen. We changed his haloperidol to risperidone 0.5mg at night, with a view to decrease EPSE, and trihexyphenidyl was stopped. Subsequently, he was weaned off the catheter successfully. Limb tremors were still significant.

In summary, according to the consensus diagnostic criteria in the DLB consortium in 2005,¹⁰ the patient was diagnosed with probable DLB, supported by fluctuating, vivid, visual hallucinations and increased neuroleptic sensitivity. He was diagnosed with Meige syndrome in 1999 (i.e., 11 years before he developed DLB).

DISCUSSION

The neurological diseases associated with LB form a disease spectrum¹¹ based upon the number of LB, distribution of LB, and associated neuronal loss. LB is detected in the brains of about 10 percent of clinically normal, elderly people over the age of 60. When LB is found in normal individuals, it is

sometimes referred to as incidental Lewy body disease, which remains asymptomatic due to subthreshold pathology.¹² If LB involves more widespread areas in the brain, it can give rise to Parkinson's disease, DLB, and primary autonomic dysfunction. In clinical practice, patients often have heterogeneous combinations of parkinsonism, dementia, and autonomic dysfunction, reflecting pathological involvement at multiple locations.¹³ LB has also been described in many other clinical syndromes, including progressive supranuclear palsy, motor neuron disease, corticobasal degeneration, neuroaxonal dystrophy, and rapid eye movement sleep behavior disorders.¹³ Meige syndrome may represent another similarly uncommon clinical disorder that is associated with LB.

Dopamine depletion in the basal ganglia has been linked to blepharospasm in rodent models.¹⁴ Should this model apply to humans, the basal ganglia dysfunction in some Parkinson's disease or DLB cases at onset may be so mild as to express as blepharospasm, and this basal ganglia dysfunction may also explain why patients with Meige syndrome are more prone to develop parkinsonian symptoms even without prior exposure to dopamine blockers.¹⁵ As LB progressed and involved more areas in the brain, our case experienced cognitive deterioration, visual hallucinations, and sensitivity to neuroleptic medications. To our knowledge, this is the first case in which a patient developed DLB well after the diagnosis of Meige syndrome.

Nearly 20 years ago, Meige syndrome was first suggested for inclusion in the spectrum of Lewy body disease. However, there remains no definitive conclusion. The current case report describes a patient with Meige syndrome subsequently developing DLB, and illustrates the possible link between Meige syndrome and Lewy body disease.

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