Variability in the Duration of the Onset of Idiopathic Parkinson’s Disease after Essential Blepharospasm

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Dear Editor

Blepharospasm is a focal cranial dystonia characterized by excessive involuntary closure of the eyelids. Essential blepharospasm (EB), the most common form of this condition, generally occurs between the fifth to seventh decades of life and affects women more often than men (1).

Similar to EB, the prevalence of idiopathic Parkinson’s disease (IPD) increases with age. Moreover, EP has been reported to occur in a variety of neurodegenerative disorders, including Parkinson’s disease (PD) (2). Interestingly, EB does not typically precede the symptoms of Parkinsonism and is not often seen in IPD cases (1). Herein, we present two patients diagnosed with EB who later on developed IPD. Interestingly enough, the duration of the onset of IPD after EB varied significantly in both cases.

A 54-year-old male diagnosed with EB two years ago, developed a resting tremor of his left upper extremity over several days. He reported no noticeable slowness in his daily activities and no significant problems with his manual dexterity, walking, or balance. No response of tremor to alcohol was reported, and there was no history of antipsychotic drug use, metoclopramide, or exposure to toxins. The patient did not smoke or drink and had no family history of neurological disorders. The only medication he was receiving was botulinum toxin A injections, every 3-4 months, for blepharospasm.

Upon examination, he had intermittent spasmodic tonic contractions of the orbicularis oculi on both sides. Motor testing showed mild rigidity of left upper extremity and mild bradykinesia upon finger tapping, fist clenching and rapid turning of his left hand. He had moderate amplitude, 5 Hz supination-pronation resting tremor of his left upper extremity. Examination of the gait showed a slightly decreased left arm swing. Rest of the neurological examination and brain imaging was normal. A diagnosis of IPD was made.

The second patient was a 73 year-old right handed female diagnosed with EB 18 years ago, developed a resting tremor of both upper and lower extremities over a period of several days. She reported significant slowness in performing her daily activities. There was no problem with here balance. No response of tremor to alcohol was reported, and there was no history of antipsychotic or any other drug use. The only medication she was receiving was botulinum toxin A injections, every 3-4 months, for blepharospasm. Rest of history was unremarkable.

Upon examination, she had intermittent spasmodic tonic contractions of the orbicularis oculi on both sides. Motor testing showed moderate rigidity of both upper extremities, right much greater than the left side. There was mild bradykinesia upon finger tapping, fist clenching and rapid turning of both hands, right much greater than the left side. She had moderate amplitude, 5 Hz supination-pronation resting tremor of both upper and lower extremities, with the right side much greater than the left side. Examination of the gait showed a mildly decreased right arm swing. Rest of the neurological examination and brain imaging was normal. A diagnosis of IPD was made.

Dystonia and Parkinsonism are common movement disorders associated with basal ganglia dysfunction. Although dissimilar in their clinical manifestations, they share several characteristics: symptoms may be induced by dopaminergic antagonists, patients show some benefit with anticholinergic drugs, and both conditions can be caused by similar lesions to the brain (3).

There is also evidence suggesting that both environmental factors and genetic predisposition may play a role in the development of IPD and EB (1). Although our patients had no family history of neurodegenerative disorders, they may have inherited an
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autosomal dominant gene mutation, which is common in
IPD patients. This mutation could have been triggered
by an environmental stimulus and caused his medical
conditions (4). However, both of our patients developed
relatively subacute onset of IPD, which was tremor
dominant and the duration of the development of PD
was quite variable in both cases. It has been suggested
that increased inhibitory output at the basal ganglion’s
substantia nigra pars reticulata creates a “permissive”
condition in the trigeminal sensory-motor blink circuits
that could result in EB in rodent models. Should this
hypothesis apply to humans and the pathophysiological
mechanisms are similar in both cases reported, the
question arises why there was a significant variability in
the duration of the onset of IPD followed by EB in both
cases (5).

Overall, it is not known how long nigral
degeneration begins before the signs of PD are observed.
Results from clinical, pathological, and imaging studies
suggest that a period of 6-8 years is likely, but probably
longer in patients with genetic causes of PD (6). Further
research into biomarkers and more accurate imaging
tools can perhaps uncover the pathological correlations
between IPD and EB. Patients with EB should be
periodically monitored for signs of IPD.

References

1. Defazio G, Livrea P. Primary blepharospasm: diagnosis
and management. Drugs 2004;64(3):237-44.
2. Ben-Shlomo Y, Sieradzan K. Idiopathic Parkinson's
disease: epidemiology, diagnosis and management. Br J
3. Micheli F, Scorticati MC, Folgar S, Gatto E. Development
of Parkinson's disease in patients with blepharospasm.
Mov Disord 2004;19(9):1069-72.
CW. Altered proteosomal function in sporadic Parkinson's
5. Micheli F, Scorticati MC, Folgar S, Ge. Development of
Parkinson's disease in patients with blepharospasm. Mov
Disord 2004; 19(9):1069-72.
6. Schapira AH. Neurobiology and treatment of Parkinson's