

### In Response To:

Onder H. Spontaneous intracranial hypotension and its association with movement disorders? Tremor Other Hyperkinet Mov. 2016; 6. doi: 10.7916/D84B31NS

**Original Article:** 

Salazar R. Spontaneous intracranial hypotension associated with kinetic tremor and ataxia. Tremor Other Hyperkinet Mov. 2016; 6. doi: 10.7916/D8HQ3ZN5

## Letters

## Reply to: Spontaneous Intracranial Hypotension and its Association with Movement Disorders?

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I thank Dr. Onder for raising important points related to this case report.<sup>1</sup> Certainly the identification of the cerebrospinal fluid (CSF) leak would have enabled us to perform a target-oriented therapy (i.e., epidural blood patch), and an adequate clinical response further supported the diagnosis of spontaneous intracranial hypotension (SIH). That being said, the identification of a CSF leak is not necessary to fulfill the diagnostic criteria for this syndrome.<sup>2</sup> Other studies have reported cases of SIH where the CSF leak could not be identified after thorough investigation.<sup>3</sup> Along those lines, treatment with an epidural blood patch is the recommended therapy for patients who fail to respond to conservative measures. In this case, SIH-associated movement disorders, namely gait ataxia, kinetic tremor and gait start hesitation, were managed with conservative measures including adequate hydration, bed rest, and increased caffeine consumption. At the 6-month follow-up appointment, his physical examination demonstrated improvement in gait ataxia and tandem gait without significant change in the degree of kinetic tremor or intermittent gait

start hesitation. More importantly, no new neurological signs or symptoms were recognized after 3 years from symptom onset. Overall, the patient was satisfied with the clinical response. Unfortunately, no follow-up neuroimaging study was performed because the patient did not keep any further appointments following the initial good response.

I agree with the importance of assessing long-term clinical outcomes of conservative or surgical intervention on SIH-associated movement disorders, especially since the pathophysiology of these symptoms remains poorly understood. The most accepted theory of the pathophysiologic mechanism of SIH headaches and other associated neurological symptoms (i.e., diplopia, facial numbness, dizziness, radicular symptoms) proposes a decrease in CSF volume that causes a subsequent reduction in the buoyant force, leading to traction of several pain-sensitive structures. However, this theory fails to explain the expanding clinical spectrum of SIH reported in case studies and case series that consist of ataxia, chorea, tremor, dystonia, and parkinsonism.<sup>1,3</sup>

This case further illustrates the spectrum of movement disorders associated with SIH. Needless to say, the differential diagnosis of an older patient presenting with chronic parkinsonism and ataxia should preferentially include neurodegenerative disorders like multiple system atrophy, fragile X-associated tremors and ataxia, among others. Being a movement disorders specialist, I completed consecutive physical exams on this patient to scrutinize any signs suggestive of an underlying Parkinson plus syndrome or other neurodegenerative disorder. The last physical exam was performed approximately 3 years after disease onset and demonstrated the absence of additional secondary signs (i.e., dysarthria, dysautonomia, cognitive decline, bulbar dysfunction, rigidity, or bradykinesia). Nevertheless, I concur with Dr. Onder's appraisal that the diagnosis of an uncommon clinical presentation of SIH with associated movement disorders should be contemplated as a diagnosis of exclusion after the completion of a thorough investigation of more common neurodegenerative disorders. Larger studies are necessary to understand the pathophysiological mechanisms of movement disorders associated with SIH. I hope this case report stimulates others to report similar uncommon presentations associated with SIH.

#### References

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