

Spinocerebellar ataxia type 2 in India, response to Yoo *et al.* Spinocerebellar ataxia type 2 presenting with chorea: Korean cases

We appreciate article by Yoo *et al.*¹ describing two Korean patients of spinocerebellar ataxia (SCA) type 2 presenting with chorea. Movement disorders can be one of the prominent non-ataxic features in SCA patients, and even precede onset of ataxia. Though Chorea has been reported frequently from western populations with SCA 2, the literature from Asian population is sparse.

SCA 2 is the most common type of SCA in India.² We would like to add our observation on Indian cohort of SCA 2 patients. Wadia *et al.* from Mumbai, India; in their study observed chorea in 4 out of 53 SCA 2 patients from 31 families.³ These families belonged to geographically distinct locations in the country. Bhalsing *et al.* reported chorea in a 38-year-old lady who was SCA 2 positive from southern part of India.⁴ In Another study from Bangalore, southern part of India, chorea was reported in 2 out of 28 (7.1%) patients with SCA2.⁵ However, in their study from northern part of India, Radhakrishnan *et al.* reported that none had chorea among 34 unrelated SCA 2 patients.⁶ But other movement disorders like cervical dystonia (n=2), action tremors of hand (n=4), neck tremor (n=2) and parkinsonism (n=1) were observed. In one patient; onset of cervical dystonia preceded the onset of ataxia.

Like Huntington's disease and other phenocopy syndromes (SCA 17 and dentatorubral-pallidoluysian atrophy), SCA 2 is also a polyglutamate disorder characterized by CAG repeat expansion.⁷ The various manifestations of SCA 2 may be due to presence of neuronal intranuclear inclusions at various locations.⁴

The presence of particular movement disorder in SCA may help to direct further genetic tests. In future, when encountered with a patient of chorea with mild/ no ataxia, one should consider the possibility of SCA 2.

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REFERENCES

1. Yoo D, Lee JY, Jeon BS. Spinocerebellar ataxia type 2 presenting with chorea: Korean cases. *Neurol Asia* 2018; 23: 361-2.
2. Sinha KK, Jha DK, Sinha S Spinocerebellar Ataxia (SCA 2) is the commonest among autosomal dominant cerebellar ataxia India. *Ann Indian Acad Neurol* 2000; 3:128
3. WadiaN, Pang J, Mankodi A, Desai M, Chamberlain S. A clinicogenetic analysis of six Indian spinocerebellar ataxia (SCA2) pedigrees. The significance of slow saccades in diagnosis. *Brain* 1998; 121:2341-55.
4. Bhalsing KS, Sowmya V, Netravathi M, Jain S, Pal PK. Spinocerebellar Ataxia (SCA) type 2 presenting with chorea. *Parkinsonism Relat Disord* 2013;19: 1171-2.
5. Jhunjhunwala K, Netravathi M, Purushottam M, Jain S, Pal PK. Profile of extrapyramidal manifestations in 85 patients with spinocerebellar ataxia type 1, 2 and 3. *J Clin Neurosci* 2014; 21:1002-1006.
6. Radhakrishnan DM, Goyal V, Srivastava AK, Shukla G, Behari M. Evaluation of various movement disorders in patients of genetically proven spinocerebellar ataxia: A study from a Tertiary Care Center in Northern India. *Ann Indian Acad Neurol* 2018; 21:24-8.
7. Andrew SE, Goldberg YP, Kremer B, *et al.* Huntington disease without CAG expansion: phenocopies or errors in assignment? *Am J Hum Genet* 1994; 54: 852-63.