

Prevalence and Spectrum of Non-Motor Symptoms in Patients of Spinocerebellar Ataxias in Kashmiri Population

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ABSTRACT

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Background: Spinocerebellar ataxias (SCAs) are progressive neurodegenerative disorders increasingly recognized to have substantial non-motor manifestations. Data regarding these symptoms in the Kashmiri population remain limited.

Objective: To assess the prevalence and pattern of non-motor symptoms among genetically confirmed SCA patients and compare them with healthy controls.

Methods: This observational cross-sectional study was conducted from December 2021 to December 2023 at the Government Super Speciality Hospital, Srinagar. Twenty-five clinically and genetically confirmed SCA patients and 26 age- and sex-matched healthy controls were included. Anxiety, depression, fatigue, sleep disturbances, pain, and cognition were assessed using validated questionnaires. Disease severity was assessed using the Scale for Assessment and Rating of Ataxia (SARA). Data were analyzed using the Mann-Whitney test, with $p \leq 0.05$ considered statistically significant.

Results: Among 25 patients, SCA1 was the most common subtype (52%), followed by SCA2 (40%). The median SARA score was 12 (5-37). Compared with controls, SCA patients had significantly higher median scores for anxiety, depression, fatigue, REM sleep behavior disorder, daytime sleepiness, and pain, and significantly lower MMSE scores (all $p < 0.05$), except for restless legs syndrome. Fatigue was significantly greater in SCA2 than SCA1 (median FSS 4.0 vs. 2.7; $p = 0.005$). Pain was reported by 19 patients.

Conclusion: Non-motor symptoms are common among genetically confirmed SCA patients and represent an important component of disease burden. Routine assessment and early management of these manifestations should complement evaluation of motor symptoms.

Keywords: Ataxia, REM Sleep Behaviour disorder, Anxiety, Depression, Fatigue

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INTRODUCTION

Spinocerebellar ataxias (SCAs) are a genetically varied collection of progressive disorders that are inherited in an autosomal dominant manner. Clinically, they are marked by a deterioration in balance and coordination, along with slurred speech, typically beginning in adulthood. From a genetic standpoint, SCAs are divided into those caused by repeat expansions, like SCA3/Machado–Joseph disease (MJD), and less common SCAs that arise from non-repeat mutations, such as SCA5 [1]. Most mutations associated with SCAs cause considerable harm to cerebellar Purkinje neurons, leading to cerebellar atrophy. Additionally, other parts of the nervous system, including the spinal cord, basal ganglia, and pontine nuclei of the brainstem, may also be affected.

Epidemiological studies indicate that the prevalence of spinocerebellar ataxias (SCAs) varies considerably worldwide, with most population-based studies reporting prevalence estimates of approximately 1–5.6 cases per 100,000 population. This marked geographic variability is largely explained by founder effects, ethnic differences, and variations in study methodology [2]. A nationwide study in Cuba reported a high prevalence of 8.91 cases per 100,000 inhabitants, with SCA2 being the most common subtype [3].

Currently, there are numerous genetically distinct forms of SCAs, primarily SCA 1–48, with loci exhibiting tandem repeat nucleotide expansion representing the more prevalent types of ataxias (SCA 1, SCA2, SCA 3, and SCA 6 globally). Across Asia, the reported prevalence is less consistent owing to variability in genetic testing accessibility, but common SCAs (SCA1, SCA2, SCA3, and SCA6) are predominant [4]. In India, the most frequently reported type of SCA is SCA 12 (8.6%), followed by SCA 2 (8.5%), SCA 1 (4.8%), SCA 3 (2%), SCA 7 (0.5%), and SCA 6 (0.1%) [5].

The non-motor symptoms (NMS) of traditional motor-predominant disorders have garnered significant interest in recent years. It is commonly accepted that cerebellar function is primarily associated with motor control. However, over the past few decades, there has been increasing debate regarding the non-motor functions of the cerebellum. Clinical neuroimaging, anatomical connectivity, and neuropsychological studies have demonstrated the existence of cerebello-cerebral neural circuits involved in cognition and emotion, which are disrupted in cerebellar disorders [6]. These findings may offer additional insights into the mechanisms underlying cerebellar cognitive affective syndrome, a term coined by Schmahmann, and characterized by executive dysfunction, impaired spatial cognition, personality changes, and linguistic difficulties. Recent research has revealed that NMS are more widespread than previously thought, not only in various neurodegenerative disorders like synucleinopathies (including Parkinson's disease, Lewy body dementia, and multiple system atrophy), tauopathies (such as Alzheimer's disease, frontotemporal dementia, corticobasal degeneration, and progressive supranuclear

palsy), amyotrophic lateral sclerosis, and prion diseases, but also in spinocerebellar ataxia (SCA). These symptoms can cause significant distress to patients and their caregivers and pose therapeutic challenges [7].

We hypothesized that patients with genetically confirmed spinocerebellar ataxias (SCAs) in Kashmiri population exhibit a high prevalence and distinct patterns of non-motor symptoms (NMS) compared to healthy controls, with specific SCA subtypes demonstrating unique non-motor symptom profiles. Identifying these differences within our population using a set of questionnaires will underscore the need for subtype-specific clinical assessment and comprehensive management strategies that address both motor and non-motor manifestations. The Kashmiri population, characterized by relative genetic homogeneity, geographical clustering, and higher prevalence of consanguinity, offers an opportunity to explore disease patterns, especially in the context of the sparsity of regional data.

MATERIALS AND METHODS

Study Design: Observational cross-sectional study was approved by the local institutional ethics committee (Ref. No: IRBGMC/C5-8)

Research Location: This study took place in the Neurology Department at the Government Super Speciality Hospital, affiliated with the Government Medical College in Srinagar, Jammu and Kashmir, India.

Research Period: This study conducted for 2 years from December 2021 to December 2023.

All participants provided written informed consent. The study encompassed all patients who had both clinical and genetic confirmation of SCA. For comparison purposes, an equal number of healthy controls, matched by age and sex, were also included in the study.

Sample size and recruitment of participants: Spinocerebellar ataxias are rare neurodegenerative disorders, and the availability of genetically confirmed cases is limited, particularly in a single-centre setting. Therefore, a formal a priori sample size calculation was not feasible. Therefore, this study adopted a consecutive sampling strategy, wherein all eligible patients with clinically and genetically confirmed spinocerebellar ataxia presenting to the Department of Neurology during the study period were screened and recruited. A group of 25 patients who met the specified inclusion and exclusion criteria and gave their informed consent were included in the study. For comparison purposes, 26 healthy controls, matched by age and sex, were selected through purposive sampling from hospital staff and community volunteers, ensuring they had no neurological or systemic illnesses. Given the rarity of spinocerebellar ataxias, the sample size was determined by the number of eligible patients available during the study period. This pragmatic approach is consistent with methodological recommendations for clinical research in rare diseases, where

feasibility constraints frequently influence the achievable sample size [8].

Inclusion Criteria: All patients had definite clinical and genetic diagnoses of SCA.

Exclusion Criteria: Patients with concomitant disorders, such as diabetes, COPD and coronary artery disease.

Once consent was obtained, patients participated in interviews using a pre-designed questionnaire, and their baseline demographic and clinical information was documented. Patient age, age at onset, duration of illness, sex, and educational status were recorded. Family history of ataxia, type of inheritance, number of family members affected, and presence of other neurodegenerative diseases in the family were inquired. A detailed neurological examination, with special emphasis on cerebellar signs, was performed. The Scale for Assessment and Rating of Ataxia (SARA) score was utilized to evaluate the severity of ataxia at the time of presentation.

All participants were interviewed for non-motor symptoms (NMS), such as depression, anxiety, sleep disturbances, fatigue, and pain, using validated questionnaires. These included Hamilton Anxiety Rating Scale and Hamilton Depression Rating Scale (HAM-A and HAM-D) for anxiety and depression [9]. Recent studies have demonstrated that the Hamilton Depression Rating Scale (HDRS) maintains good criterion validity against DSM-based diagnostic interviews, with strong cross-cultural applicability across multiple language versions [10,11]

Various questionnaires were used to assess sleep disturbances. REM sleep behavior disorder was screened using the REM Sleep Behavior Disorder Screening Ques-

tionnaire (RBDSQ), a validated instrument with good reliability and diagnostic accuracy across multiple populations and language adaptations [12]. Restless legs syndrome severity was assessed using the International Restless Legs Syndrome Rating Scale (IRLS), a validated 10-item clinician-administered instrument developed by the International Restless Legs Syndrome Study Group, with established reliability, validity, and responsiveness for assessing disease severity [13]. Daytime sleepiness was assessed using the Epworth Sleepiness Scale (ESS), a widely used self-administered instrument originally described by Johns (1991), with subsequent studies demonstrating acceptable reliability, validity, and cross-cultural applicability [14,15].

Fatigue severity was assessed using the Modified Fatigue Severity Scale (FSS), a widely used instrument originally described by Krupp et al. (1989), with subsequent studies demonstrating good reliability, validity, and applicability in neurodegenerative disorders, such as Parkinson's disease [16,17]. The Brief Pain Inventory (BPI) was used to assess pain intensity and its interference with daily activities. Originally developed by Cleeland and Ryan, the BPI is a widely used multidimensional pain assessment instrument with established reliability, validity, and applicability across diverse clinical populations, including patients with chronic musculoskeletal pain and neurological disorders [18].

The Mini-Mental State Examination (MMSE) developed by Folstein was utilized to evaluate cognitive abilities.

Control subjects were interviewed for the HAM-A, HAM-D, RBDSQ, RLS Questionnaire, ESS, FSS, and MMSE.

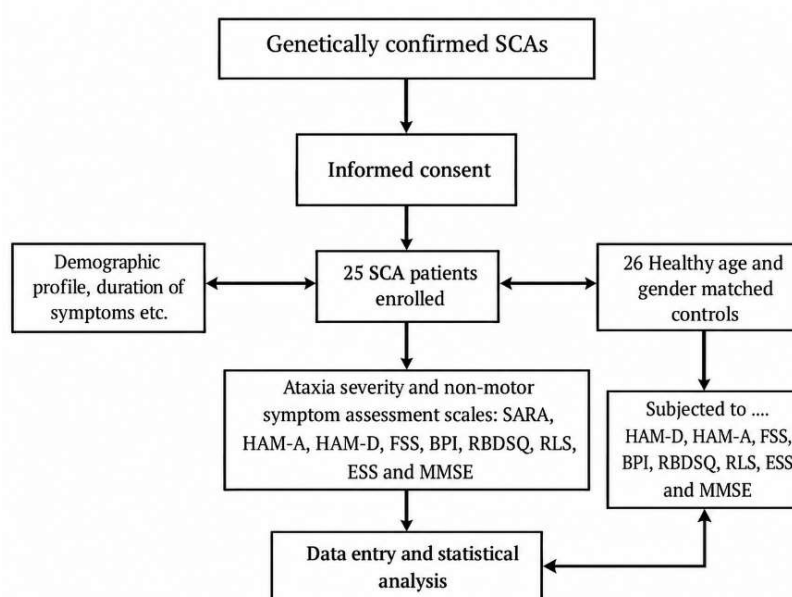


Figure 1: Flow chart of study enrollment and assessment protocol

Statistical analysis: The data were processed using SPSS software version 20. Descriptive statistics were

employed to present the data, with continuous variables shown as medians along with their maximum and mini-

mum values, and categorical variables represented by frequencies and percentages. For continuous variable analysis, the Mann-Whitney test, a non-parametric method, was applied. A p-value of ≤ 0.05 was deemed significant.

Ethical Approval: The study was approved by the Institutional Ethical Review Board of Government Medical College, Srinagar (Ref. No. IRBGM/C5-8, dated March 8, 2022).

RESULTS

Demography: The study included 25 (13 males and 12 females) genetically confirmed autosomal dominant SCA patients (13 SCA1, 10 SCA2, 1SCA3 and 1SCA12). The patients were a median age of 40 years, with ages ranging from 18 to 65 years. The age, sex distribution, and SCA type are shown in table 1,2 and 3, respectively.

The combined median SARA score was 12 (5 - 37). There were 26 age- and sex-matched controls (13 males and 13 females) with a median age of 40 (18 - 65). The details are presented in Table 4.

The SARA score was higher in SCA type 2 than in SCA type 1. The highest SARA score was 37, which was observed in SCA type 3. The SARA score increased with an increase in the duration of symptoms in a particular SCA subtype.

Depression and anxiety in SCA: The median HAM-D score was 5 (2-16). Three patients had scores of ≥ 8 , suggestive of mild depression (two patients with SCA2 and one patient with SCA1), and one patient had a score >14 , suggestive of moderate depression (one SCA3 patient). The median HAM-A score was 6 (range, 1-16).

Fatigue in SCA: Most of the patients in our study group complained of significant fatigue, with a median FSS score of 3.3 (1.7-4.5).

Sleep disturbances in SCA: Three patients had an RBDSQ score of ≥ 6 , suggestive of RBD, with a median RBDSQ score of 2 (0-6). All three patients with RBD had SCA2. Four patients were found to have RLS, with a median RLS score of 0 (0-24). All four patients had minimal RLS symptoms. Among these four patients, three had SCA2 and one had SCA1. The median ESS score was 7 (0-11).

Pain in SCA: Nineteen patients complained of significant pain, with a median BPI score of 3.5 (0-5). Most of these

Table 1: Age distribution of cases

Age group	Number (%)
16-30	8 (32)
31-45	9 (36)
46-60	5 (20)
61-75	3 (12)
Total	25 (100)

Table 2: Gender distribution of cases

Gender	Number (%)
Male	13 (52)
Female	12 (48)
Total	25 (100)

Table 3: Distribution of SCA type among the cases

SCA type	Number (%)
SCA type 1	13 (52)
SCA type 2	10 (40)
SCA type 3	1 (4)
SCA type 12	1 (4)
Total	25 (100)

patients had pain in multiple regions of the body, mostly musculoskeletal pain.

MMSE in SCA: The median MMSE score for the entire group was 27 (19-30), with a minimum score of 19. The lowest MMSE score was observed in SCA3.

SCA1 vs SCA2: The median scores of different non-motor scales were compared between SCA1 and SCA2 patients. The median FSS score difference between the two groups was statistically significant ($p = 0.005$), suggesting significant fatigue in SCA2 patients compared with SCA1 patients (Table 5). The prevalence of RBD and RLS was similar in SCA1 and SCA2, with no statistically significant difference. The observed findings were based on the scores and symptom profiles recorded in the study, with the subtype comparison focused specifically on SCA1 and SCA2 patients for the relevant non-motor scales assessed.

Non-motor symptoms were compared between SCA (combined) patients and healthy controls. The difference in median scores for all non-motor scales (except RLS) between the two groups was statistically significant, indicating a higher burden of non-motor symptoms in the SCA group.

Table 4: Demographic characteristics of study participants

Characteristic	Healthy controls	SCA (Combined)	SCA type 1	SCA type 2	SCA type 3	SCA type 12
Total no	26	25	13	10	1	1
Gender (M/F)	13/13	13/12	7/6	6/4	0/1	0/1
Age in years*	40 (18-65)	40 (18-65)	32 (20-65)	41.5 (18-65)	56	45
Duration of symptoms in years*	-	4 (1-12)	4 (1.5-12)	3.5 (1-10)	6	5
SARA score	-	12 (5-37)	10 (9-17)	13 (5-15)	37	12

*(Median, min-max)

Table 5: Non-motor symptoms among patients with spinocerebellar ataxia

Non-motor symptom questionnaire	SCA, combined	SCA type 1	SCA type 2	p value†
HAM-A, median (min-max)	6 (1-16)	6 (1-12)	6 (2-16)	0.851
HAM-D, median (min-max)	5 (2-16)	5 (2-9)	7 (2-16)	0.064
FSS, median (min-max)	3.3 (1.7-4.5)	2.7 (1.7-3.6)	4.0 (2.2-4.5)	0.005
RBDSQ, median (min-max)	2 (0-6)	2 (0-5)	2.5 (0-6)	0.221
RLS, median (min-max)	0 (0-24)	0 (0-22)	0 (0-24)	0.189
ESS, median (min-max)	7 (0-11)	6 (0-11)	7 (3-11)	0.191
BPI, median (min-max)	3.5 (0-5)	3 (0-5)	3.75 (0-5)	0.312
MMSE, median (min-max)	27 (19-30)	28 (23-30)	26.5 (25-30)	0.900

Notes: HAM-A, Hamilton Anxiety Rating Scale; HAM-D, Hamilton Depression Rating Scale; FSS, Fatigue Severity Scale; RBDSQ, REM Sleep Behavior Disorder Screening Questionnaire; RLS, Restless Legs Syndrome scale; ESS, Epworth Sleepiness Scale; BPI, Brief Pain Inventory; MMSE, Mini-Mental State Examination.

interpretive thresholds/cut-offs: HAM-A: <17, mild anxiety; 18-24, mild-to-moderate anxiety; 25-30, moderate-to-severe anxiety. HAM-D: 0-7, normal; 8-13, mild depression; 14-18, moderate depression; 19-22, severe depression; >23, very severe depression. FSS ranges from 1-7, with higher scores indicating greater fatigue. RBDSQ score ≥ 6 is suggestive of REM sleep behavior disorder. RLS ranges from 0-40, with higher scores indicating greater severity. ESS ranges from 0-24, with higher scores indicating greater daytime sleepiness. BPI ranges from 0-10, with higher scores indicating greater pain severity. MMSE: 24-30, no cognitive impairment; 18-23, mild cognitive impairment; <17, severe cognitive impairment. †p value for comparison between SCA type 1 and SCA type 2.

Table 6: Comparison of non - motor symptoms among cases and controls

Non-motor symptom questionnaires	SCA (Combined)	Controls	p value
HAM – A (Median, Min-Max)	6 (1-16)	1 (0-7)	<0.001
HAM – D (Median, Min-Max)	5 (2-16)	2 (0-6)	<0.001
FSS (Median, Min-Max)	3.3 (1.7-4.5)	1 (1-2)	<0.001
RBDSQ (Median, Min-Max)	2 (0-6)	0.5 (0-3)	0.027
RLS (Median, Min-Max)	0 (0-24)	0 (0-8)	0.13
ESS (Median, Min-Max)	7 (0-11)	4 (1-9)	0.023
BPI (Median, Min-Max)	3.5 (0-5)	0.5 (0-3.7)	0.001
MMSE (Median, Min-Max)	27 (19-30)	30 (26-30)	<0.001

*MMSE, Mini-Mental Status Examination; FSS, Fatigue Severity Scale; HAM-D, Hamilton Depression Rating Scale; HAM-A, Hamilton Anxiety Rating Scale; RLS, restless leg syndrome; ESS, Epworth Sleepiness Scale; RBD, rapid eye movement sleep behavior disorder; BPI, pain inventory.

DISCUSSION

In this cross-sectional study of genetically confirmed SCAs from the Kashmiri population, we observed a high prevalence of non-motor symptoms compared to healthy controls, with prominent fatigue, sleep-related symptoms, pain, and lower cognitive screening scores. These findings support the need for routine screening for non-motor manifestations alongside motor severity assessment in clinical care.

The cerebellum has long been recognized as a crucial component of the motor system, playing a key role in movement coordination and motor learning. However, recently interest has shifted towards its involvement in non-motor functions. This shift is highlighted by the "dysmetria of thought" hypothesis and the clinical identification of cerebellar cognitive affective syndrome (CCAS), which is characterized by executive dysfunction, challenges in spatial cognition, language difficulties, and affective dysregulation. Its validity and clinical significance have been reinforced by subsequent meta-analytic and consensus-based studies [19].

Our cohort comprised 25 patients with genetically confirmed SCA, with SCA1 as the most common subtype (52%, n = 13), followed by SCA2 (40%, n = 10); one patient each had SCA3 and SCA12, respectively. Mastammanavar VS et al. [7] conducted a similar study on non-

motor symptoms in autosomal-dominant SCA, including 41 patients (SCA1, n = 17; SCA2, n = 14; SCA3, n = 10).

The median age of our cohort was 40 years (range, 18-65 years), with males constituting 52% (n = 13). In the study by Mastammanavar et al. [7], the combined mean age was 32.9 ± 9.2 years, and males constituted 66% (n = 27).

The median SARA score in our study was 12, with a maximum score of 37, which was seen in 1 SCA type 3. The SARA score was higher in SCA type 2 than in SCA type 1.

Depression and anxiety in SCA: Depression has been well established in SCA subtypes. It has been described in SCA1, SCA2, SCA3, and SCA6; however, it is predominantly seen in SCA3 patients (33.5-60%). Depression was observed in four patients in our study, with mild depression in three patients (two SCA2 and one SCA1 patient). We had only one SCA3 patient in our study whose HAM-D score was 15, suggesting moderate depression. The median HAM-D score was higher in patients with SCA than in healthy controls, and this difference was statistically significant (p < 0.00).

In the study by Mastammanavar VS et al. [7], more than half of the patients had depression (n = 25, 60.97%). The combined mean HAM-D score was 10.7 ± 11.0 . Pa-

tients with SCA3 had higher HAM-D scores (13.07 ± 13.9).

In a recent cohort study conducted in the Thai population by Jekpoo et al., approximately 27% of patients with SCA exhibited clinically significant depression, with symptom severity strongly correlating with anxiety, stress, and disease-related psychosocial factors [20]. These findings underscore the contribution of cerebellar–cortical network dysfunction to affective disturbances and highlight the need for routine neuropsychiatric assessment in patients with SCA.

In our study, the median HAM-A score was higher in patients with SCA than in healthy controls, and the difference was statistically significant. Mastammanavar VS et al. [7] in their study also reported significant anxiety symptoms in patients with SCA. Recent evidence highlights the cerebellum as a key modulator of cognitive and affective functions through its extensive connections with distributed cortical networks, extending its role beyond motor control [21].

Sleep disturbances in SCA: Sleep disturbances are increasingly recognized as important non-motor manifestations of spinocerebellar ataxia (SCAs) types 1, 2, 3, and 6. Studies have consistently shown a higher burden of sleep complaints than that in healthy controls. A cross-sectional cohort study of patients with spinocerebellar ataxia type 3 reported that sleep disturbances are among the common non-motor manifestations, supporting the need for routine screening in clinical practice [22]. Contemporary reviews of non-motor features in SCAs similarly highlight sleep disturbance as a frequent contributor to reduced quality of life.

In our study, the median RBDSQ and ESS scores were higher in patients with SCA than in healthy controls, and the difference was statistically significant ($p < 0.05$). Sleep disturbances are more common in the SCA3 subtype. However, subtype comparisons in our cohort were limited by the small sample size. Although not statistically significant, we observed that sleep-related symptoms were more frequent in SCA2 than in SCA1.

Fatigue in SCA: Fatigue is another common symptom that is under-recognized in patients with SCA. In our study, it was observed in almost all SCA groups but was more prevalent in SCA2. The median FSS score was higher in SCA2 than in SCA1, and this difference was statistically significant ($p < 0.005$). Brusse et al. found fatigue to be one of the most common disabling symptoms in patients with SCA, particularly in SCA3 [23]. Physical and mental functioning, sleep, and depression were the strongest fatigue predictors. When evaluating treatment strategies, it is crucial to tackle all controllable factors contributing to fatigue, with particular emphasis on addressing depression, vision problems, and sleep disturbances.

Pain in SCA: Pain was observed in 19 of the 25 patients in our study. The most common symptoms were musculoskeletal pain and backache. The median BPI score

was higher in SCA2 (3.75) than in SCA1 (3.0), although this difference was not significant. Neurophysiological studies on spinocerebellar ataxias have demonstrated widespread abnormalities in peripheral nerve excitability and motor neuron function, reflecting involvement beyond cerebellar pathways and supporting a multisystem neurodegenerative process underlying clinical manifestations, such as muscle cramps and motor dysfunction [24].

Cognitive dysfunction in SCA: The median MMSE score of our patients was 27 (19–30) and was significantly lower than that of healthy controls [median MMSE of 30 (26–30)]. This difference was statistically significant ($p < 0.00$). The lowest MMSE score (19) was observed in one SCA3 patient. Cognitive impairment has been described in various SCA types. This has been postulated to be due to the disruption of the cerebro-cerebellar connections at various levels. Mastammanavar VS et al. [7] reported significant cognitive impairment in SCA patients compared to healthy controls.

The cerebellar vermis is considered a limbic region involved in emotion. The lateral cerebellar hemisphere is associated with cognition. The right posterior cerebellum is associated with language function, whereas the left cerebellar hemisphere is associated with visuospatial functions. Executive functions are subserved by both hemispheres.

LIMITATIONS

Our study had a few limitations. This was a single-center study with a small sample size. The sample size was neither adequate nor uniform across the groups. Our study was a cross-sectional study with no follow-up. Non-motor manifestations were assessed using questionnaires, and the symptoms may vary over time.

CONCLUSION

Our research highlights that non-motor symptoms are common and negatively impact the quality of life for individuals with SCAs. Future studies are needed to explore whether these extracerebellar symptoms develop before ataxia and the pathophysiological mechanisms involved in the development of these symptoms. It is crucial to identify the non-motor symptoms of SCAs and begin treatment early in the disease course.

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Individual Author's Contribution: BAS contributed to the

conception and design of the study, data collection, and data analysis and interpretation. **SAT** contributed to the conception and design of the study, data collection, data analysis and interpretation, and manuscript preparation. **TH** contributed to the conception and design of the study, data collection, data analysis and interpretation, and manuscript preparation. **RQ** contributed to study design, data analysis and interpretation, and manuscript preparation. **SHA** contributed to the conception and design of the study and data collection. **ARK** contributed to the conception and design of the study and data collection. All authors reviewed and approved the final manuscript.

Availability of data: The data that support the findings of this study are available from the corresponding author on reasonable request.

Declaration of Non-use of generative AI Tools: During the preparation of this manuscript, the author(s) used AI tool (Paperpal) solely for the purpose of improving language, grammar, and readability. The tool was not used to generate original content, data, analysis, or scientific conclusions. After using this tool, the author(s) reviewed and edited the content as necessary and take full responsibility for the content of the publication.

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