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**RESEARCH ARTICLE**

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# Retrospective Review of 56 Patients with Sensory Ganglionopathy

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## ABSTRACT

**Introduction:** Sensory ganglionopathy (SG) is a rare and specific subgroup of the peripheral nervous system diseases. Electrophysiology shows a widespread amplitude decrease in sensory nerve action potential (SNAP) or absence of SNAPs in SG. SG diagnosis has a special importance in the context of etiology. We aimed to contribute to diagnosing SG as early as possible with the help of other supportive diagnostic work-up without the need for invasive methods by reviewing data of patients with electrophysiologically diagnosed SG.

**Methods:** The medical records of 56 patients with SG diagnosed by electrodiagnostic (EDx) tests were reviewed. EDx findings were compared with our laboratory normal values. Also EDx findings of patients with 23 pure SG patients and 9 SG with motor axonal involvement were analyzed.

**Results:** Etiology was determined in 35 (62.5%) of all patients besides a considerable idiopathic preponderance (n=14; 25%). Comparison of ulnar nerve DMLs of 23 pure SG patients and 9 SG with motor axonal involvement patients, was not statistically significant (p=0.064). Needle EMG findings of lower extremity were grade 1 in 52.9% and grade 3 in 5.8%. Of the patients who underwent upper extremity needle EMG, grade 1 was detected in 53.8% and grade 3 was detected in 7.6%.

**Conclusion:** In order to diagnose SG as early as possible without the need for invasive methods and to elucidate its etiology, a thorough electrophysiological screening should be performed with the help of other supportive diagnostic tools.

**Keywords:** Distal motor latency, dorsal root ganglion, ganglionopathy, sensory neuronopathy, ulnar nerve

## INTRODUCTION

Sensory ganglionopathy (SG) or sensory neuronopathy characterized by primary and selective damage to the dorsal root ganglion (DRG) and/or sensory nuclei of the brainstem is a rare and specific subgroup of the peripheral nervous system diseases (1). Degeneration of both central and peripheral sensory projections of DRG has been established (1). Unlike other more common peripheral neuropathies, concomitant impairment of short and long peripheral axons manifests in a distinct clinical picture (2) and SGs have a common clinical and electrophysiological pattern despite diverse range of etiologies (3). Early ataxia probably due to the denervation of proximal muscle spindles and joints, deep sensory disturbance and loss of deep tendon reflexes due to the involvement of large nerve fibers, preserved strength, sensory symptoms -sometimes asymmetrically and/or patchy- at both proximal and distal sites of the body are the general clinical features of SG (1, 2).

Electrophysiology shows a widespread amplitude decrease in sensory nerve action potential (SNAP) or absence of SNAPs (1, 2). This pattern may be more pronounced in the upper extremities, and motor conduction abnormality is either absent or mild. These clinical and electrophysiological features summarized reflect nonlength-dependent pattern of peripheral axon degeneration (2) in other words axonal involvement without a distal to proximal gradient (4, 5) and it is acknowledged as the hallmark of SG (6).

The presence of various etiologies (7), such as immune-mediated diseases, vitamin deficiencies, drug toxicity, paraneoplastic syndromes and genetic causes (5) renders the early identification of SG crucial, particularly for reversible causes (6). In case histopathological study of DRG is absent or unavailable, the electrophysiological studies are crucial for the diagnosis of SG. In such cases, electrophysiological studies and imaging methods complement each other. In fact, all efforts are aimed at distinguishing SG from sensory axonal polyneuropathy and making SG's early diagnosis.

In this study, SG is assessed primarily from a neurophysiological perspective with supportive diagnostic work-up.

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## Highlights

- **The records of 56 patients with sensory ganglionopathy (SG) were reviewed.**
- **The etiology was identified in 62.5% of patients, while 25% had idiopathic causes.**
- **Grade 1 was found in 52.9% of lower and 53.8% of upper extremity EMG examinations.**
- **For the diagnosis of SG, a thorough EDx screening is recommended.**
- **For the diagnosis of SG, the aid of other supportive diagnostic tools is important.**

# METHODS

### Subjects

Hacettepe University EMG Laboratory database was searched by "ganglionopathy" and "neuronopathy" from 2017 to 2024. The electrodiagnostic (EDx) records of matched 56 patients were evaluated according to the neurophysiological parameters of the Camdessanché criteria (3). SG diagnoses were also subsequently confirmed clinically were reviewed. The local ethics committee approved the study (Hacettepe University, No. SBA 24/196).

### Electrophysiology

### Sensory Nerve Conduction Studies

The bandpass filter was set at 20 Hz-2KHz. NCSs were performed using a Keypoint- Denmark EMG machine. Skin temperature was maintained at or above 32°C.

Median second finger and ulnar sensory NCSs were performed at least unilaterally in all patients. The stimulator was placed in the proximal sites of the second and fifth digits in median and ulnar nerve sensory NCSs, respectively. The active and reference recording cup electrodes, 3 cm apart, were placed on the wrist in the median and ulnar nerve sensory NCSs. A subset underwent superficial radial sensory NCSs in whom superficial radial sensory NCS, the active recording cup electrodes, 3 cm apart, were placed over the anatomical "snuff box". Stimulation was 10 cm from the active recording electrode, over the dorsolateral edge of the radius bone. Supramaximal stimulation of the nerves was performed.

Sural sensory NCSs were performed bilaterally in all patients. The active recording electrode was placed posterior to the lateral malleolus. The reference recording electrode was placed 3 cm distal to the active electrode. Stimulation was 12 cm proximal to the active recording electrode, over the distal posterolateral leg. Supramaximal stimulation of the nerves was performed.

### Motor Nerve Conduction Studies

The bandpass filter was set at 20 Hz-10 KHz. Nerve conduction studies (NCSs) were performed usinga Keypoint- Denmark EMG machine. Skin temperature was maintained at or above 32°C.

Standard motor NCSs included peroneal motor NCS recording at the extensor digitorum brevis, tibial motor NCS recording at the abductor hallucis, median motor NCS recording at the abductor pollicis brevis and ulnar motor NCS recording at the abductor digiti minimi muscles with belly-tendon montage. Median, ulnar, peroneal and tibial motor NCSs were performed at least unilaterally in all patients. Supramaximal stimulation of the nerves was performed.

### Needle EMG

The bandpass filter was set at 2Hz- 10KHz. Needle EMG was performed using a Keypoint- Denmark EMG machine.

Needle EMG was performed with a standard concentric needle in at least one distal muscle of the lower extremity. If abnormal findings were detected in the first muscle evaluated, this examination was extended to other distal and proximal muscles and, if necessary, to the upper and other lower extremities – if required myotomally-. Data from patients who underwent needle EMG are presented.

### SEP/Blink Reflex/HRV Studies

The bandpass filter was set at 10 Hz-3KHz. SEP was performed using a Keypoint- Denmark EMG machine. Skin temperature was maintained at or above 32°C.

Somatosensory evoked potential (SEP) study was carried out to assess the central sensory pathway impairment in a limited number of patients. SEPs were recorded by distal stimulation of the tibial nerve; spinal and cortical responses were labeled.

Blink reflex study and heart rate variability (HRV) analysis were performed in patients with facial numbness and autonomic symptoms.

The bandpass filter was set at 20 Hz-10KHz. Blink reflex study was performed using a Keypoint- Denmark EMG machine. For blink reflex studies, the supraorbital nerves were stimulated with the cathode over the supraorbital notches and recording electrodes were on both orbicularis oculi muscles. The latencies of the ipsilateral early response (R1) and bilateral late responses (R2) were recorded (limits: R1 <13 ms; R2 ipsilateral <40 ms, contralateral <42 ms; difference between sides: 1.2 ms for R1, 5 ms for R2).

The bandpass filter was set at 0.5 Hz-2KHz. HRV study was performed using a Keypoint- Denmark EMG machine. The short-term HRV variable standard deviation of the R-R interval (SDRR) was assessed over a period of 60 s of both normal (NB-SDRR) and deep breathing (DB-SDRR). If NB-SDRR>0.0019, the test was considered abnormal. At least a two-fold increase in DB-SDRR compared to NB-SDRR was considered normal.

### Other Supportive Tests

Records of patients who underwent spinal column magnetic resonance imaging (MRI) and sural nerve biopsy were reviewed.

### Statistical Analyses

Data analyses were conducted using the Statistical Package for the Social Sciences (SPSS) version 25 for Windows (IBM, Armonk, NY, USA). Descriptive statistics were reported using number and percentages for categorical variables and mean±SD or median, range (minimum and maximum) for continuous variables. Normality of continuous variables was assessed with Kolmogorov-Smirnov or Shapiro-Wilk tests. EDx findings were compared between patients with 23 pure SG patients (SG patients with no motor fiber involvement and/or additional entrapment neuropathy, but only sensory fibers were affected) and 9 SG with motor axonal involvement by Student T or Mann-Whitney U tests. Significance was established at $p<0.05$.

# RESULTS

### Clinical Features

According to Camdessanché criteria, 24 patients were in the possible SG group and 32 in the probable SG group. None of our patients had definite SG.

Etiology was determined in 35 (62.5%) of all patients (mean age±SD = 54±18.7, 25 female) besides a considerable idiopathic preponderance (n=14; 25%).

SG was associated with chemotherapy agent toxicity in 9 cases (16%), Friedreich ataxia in 6 cases (10.7%), Sjögren's syndrome in 5 cases (8.9%), cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) in 3 cases (5.3%), paraneoplastic syndrome in 3 cases (5.3%), sicca complex without fully developed Sjögren's syndrome in 1 case, Kennedy disease in 1 case, facial-onset sensory and motor neuronopathy (FOSMN) in 1 case, hereditary sensory and autonomic neuropathy II (HSAN II) in 1 case, celiac disease in 1 case and Miller Fisher syndrome (MFS) in 1 case.

During follow-up nodular sclerosing type Hodgkin lymphoma was also detected in 1 patient who was previously diagnosed with Sjögren's syndrome related SG.

Two patients in the toxic group were associated with chronic alcoholism.

Platinum analogs (carboplatin, cisplatin, oxaliplatin) in 6 patients -including carboplatin-paclitaxel combination in 1 patient-, bortezomib in 2 patients, vincristine-doxorubicin combination in 1 patient and thalidomide in 1 patient were held responsible for SG.

Anti-Hu positivity was detected in 1 patient in the paraneoplastic group. Autoantibodies against trisulfated heparin disaccharide and fibroblast growth factor receptor 3 were not examined in any idiopathic patients, since their specificity and pathogenic role are yet unclear.

A total of 8 patients did not come for the follow-up visits. This group included 1 patient with a strong suspicion of Friedreich ataxia, 2 with suspicion of malignancy, 2 with suspicion of spinocerebellar ataxia, 1 patients with other hereditary neurological diseases, and 2 patients with neurodegenerative processes.

## Neurophysiological Studies

### Sensory Nerve Conduction Studies

Numerical values of sensory NCSs are presented in Table 1 and 2. Ulnar sensory NCS in 14 patients and sural sensory NCS in 41 patients were studied bilaterally. Superficial radial sensory NCS was performed unilaterally in 5 patients out of routine protocol. SNAP could not be elicited in either lower or upper extremities in 36 patients (64.2%), and very low amplitude SNAPs were recorded in 20 patients. In 3 of the 20 patients with SNAPs, there was no SNAP in the upper extremities and low amplitude SNAPs in the lower extremities. The etiology in these patients was cisplatin toxicity, paraneoplastic syndrome and MFS. Asymmetrical SNAP abnormalities were observed in another 3 patients out of 20; 1 was in the idiopathic group, 1 was in the paraneoplastic group and 1 was associated with possible hereditary neurological disease.

### Motor Nerve Conduction Studies

Numerical values of motor NCSs are given in Table 1 and 2. There were 9 SG patients with mild motor axonal involvement findings. In accordance with the Camdessanché criteria, there was a decrease in CMAP amplitude in 1 lower extremity motor nerve in these 9 patients. In 2 of them, there was a mild conduction velocity slowing in lower extremity motor NCSs. Of these 9 patients, tibial distal motor latency was at the upper limit or slightly prolonged in 2 and peroneal distal motor latency was at the upper limit or slightly prolonged in 1. In 3 of 9 patients, tibial or peroneal minimal F latency was at the upper limit or slightly prolonged.

Ulnar nerve distal motor latencies (DMLs) were at the upper limit or prolonged in 27 of 56 patients (48.2%) (Fig. 1). The mean (±SD) and median (range values) of ulnar nerve DML of all patients were 2.75 (±0.51) and 2.8 (1.6-3.8) ms, respectively. Prolonged lower extremity distal motor latency was not observed in any of the patients with pure SG, whereas ulnar distal motor latency was prolonged in 10 of these patients.

In the comparison of ulnar nerve DMLs of 23 pure SG patients (mean±SD=2.63±0.56 ms) and 9 SG with motor axonal involvement patients (mean±SD=2.98±0.45 ms), a statistically significant difference was not detected (*p=0.064*). The median nerve was excluded from latency evaluation, because a subset had carpal tunnel syndrome.

### Needle EMG

Excluding 9 SG patients with mild motor axonal involvement findings, there were 19 patients with chronic diffuse or segmental root/anterior horn involvement. In 3 of the 7 patients with SG with diffuse chronic anterior horn involvement, the etiologies were determined as Kennedy disease, FOSMN and chronic alcoholism-cervical myelomalacia. 2 patients did not continue their follow-up visits during the investigation of a suspected hereditary neurological diseases. The remaining 1 patient was lost to follow-up at her own request. SG with lumbosacral anterior horn involvement was observed in 3 of 6 patients diagnosed with Friedreich ataxia. SG with multiple anterior horn involvement of lumbosacral segment was detected in 1 patient, but the etiology could not be elucidated.

Needle EMG findings of the patients were grouped as upper and lower extremity needle EMG (UEMG and LEMG) findings. Needle EMG was applied to the upper extremities in 13 patients and to the lower extremities in 17 patients. UEMG and LEMG findings were graded according to our laboratory's grading system (8-10). Of our patients' LEMG findings, 52.9% were grade 1 (minimal-mild), 41.1% grade 2 (moderate) and 5.8% were grade 3 (severe). Of the patients who underwent upper extremity needle EMG, grade 1 needle EMG findings were detected in 53.8%, grade 2 needle EMG findings were detected in 23% and grade 3 needle EMG findings were detected in 7.6%. While upper extremity needle EMG findings were normal in 2 of the patients, lower extremity needle EMG findings were not normal in any patient.

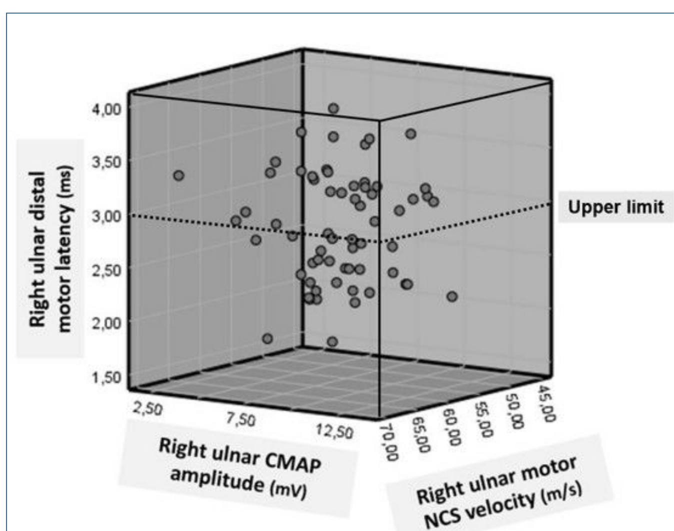


**Figure 1.** Distal motor latency/ nerve conduction study (NCS) velocity/compound motor action potential (CMAP) amplitude graph of patients' right ulnar motor nerves

**Table 1.** Numerical values of sensory and motor nerve conduction studies of patients' upper extremities

| | | | | Ulnar Nerve | | | | | | | Median Nerve | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | Sensory Nerve | | | | Motor Nerve | | | Sensory Nerve | | | | Motor Nerve | |
| | | | | Right | | Left | | | | | Right | | Left | | | |
| | n | Mean of age (SD) | Female (n) | SNAP amplitude mean(SD) (μV) | NCV mean(SD) (m/s) | SNAP amplitude mean(SD) (μV) | NCV mean(SD) (m/s) | DML mean(SD) (ms) | CMAP amplitude mean(SD) (mV) | NCV mean(SD) (m/s) | SNAP amplitude mean(SD) (μV) | NCV mean(SD) (m/s) | SNAP amplitude mean(SD) (μV) | NCV mean(SD) (m/s) | CMAP amplitude mean(SD) (mV) | NCV mean(SD) (m/s) |
| Toxic | 11 | 64.1(9.9) | 4 | 0.55(1.3) | 57(9.8) | 0 | Ø | 2.97(0.37) | 7(1.7) | 55(6.08) | 2.14(4.2) | 57.2(7.3) | Ø | Ø | 7.19(1.5) | 56.7(5.3) |
| Idiopathic | 14 | 56.09(17.3) | 4 | 0.2(0.69) | 67(0) | Ø | Ø | 2.81(0.45) | 7.7(2.2) | 55.09(3.9) | 0.43(1.4) | 61(0) | Ø | Ø | 6.45(1.4) | 54.3(2.9) |
| Friedreich ataxia | 6 | 30.1(8.2) | 4 | 0(0) | Ø | 0 | Ø | 2.45(0.42) | 7.2(2.4) | 51.8(1.4) | 0.6(1.6) | 55(0) | Ø | Ø | 6.6(1.7) | 54.5(5.8) |
| Sjögren's± Sicca syndrome | 6 | 54.6(21.2) | 4 | 0(0) | Ø | 0.56(0.98) | 66(0) | 2.63(0.58) | 7.5(1.4) | 55.6(3.01) | 0(0) | Ø | 0(0) | Ø | 6.5(3.8) | 53.2(2.8) |
| CANVAS | 3 | 59.3(5.7) | 3 | 1.2(2.1) | 52(0) | Ø | Ø | 2.76(0.55) | 6.7(3.9) | 59.3(8.3) | 1.3(2.2) | *35(0) | Ø | Ø | 4.75(6.2) | 58(9.8) |
| Paraneo-plastic | 3 | 54.6(17.09) | 2 | 0(0) | Ø | 0(0) | Ø | 2.9(0.6) | 7.9(1.2) | 55.3(2.5) | 0(0) | Ø | Ø | Ø | 7.6(0.5) | 55(2) |
| HSAN II | 1 | 34 (0) | 0 | 0(0) | Ø | Ø | Ø | 2.5(0) | 6.5(0) | 51(0) | 0(0) | Ø | Ø | Ø | 7.7(0) | 51(0) |
| Celiac disease | 1 | 24(0) | 1 | 0(0) | Ø | 0(0) | Ø | 1.9(0) | 7.3(0) | 67(0) | 2.6(0) | 47(0) | 6.1(0) | 43(0) | 11.9(0) | 63(0) |
| Miller Fisher syndrome | 1 | 34(0) | 0 | 4.1(0) | 52(0) | 4.2(0) | 55(0) | 2.2(0) | 9.1(0) | 50(0) | 5.7(0) | 52(0) | 5.1(0) | 56(0) | 7.2(0) | 52(0) |
| Kennedy disease | 1 | 46(0) | 0 | 0(0) | Ø | Ø | Ø | 2.7(0) | 5(0) | 62(0) | 0(0) | Ø | Ø | Ø | 5(0) | 58(0) |
| FOSMN | 1 | 53(0) | 0 | 5.7(0) | 64(0) | Ø | Ø | 2.9(0) | 4.6(0) | 64(0) | 7.3(0) | 66(0) | Ø | Ø | 3(0) | 55(0) |
| All patients** | 48** | 54±18.7 | 25 | 0.82(2.04) | 56(5.9) | 0.75(1.6) | 56.6(8.6) | 2.77(0.5) | 7.1(2.05) | 55.1(5.1) | 1.36(2.8) | 56.5(8.4) | 2.8(3.2) | 49.5(9.1) | 6.7(2.1) | 55.8(5.1) |

* n: Number; SD: Standard Deviation; SNAP: Sensory Nerve Action Potential; NCV: Nerve Conduction Velocity; DML: Distal Motor Latency; CMAP: Compound Muscle Action Potential; CANVAS: Cerebellar ataxia, Neuropathy and Vestibular Areflexia Syndrome; HSAN II: Hereditary Sensory and Autonomic Neuropathy Type II; FOSMN: Facial Onset Sensory and Motor Neuronopathy; Ø: Not performed
**Data of the remaining 8 patients who did not attend follow-up visits and therefore whose etiology could not be determined were not reported in the table.

**Table 2.** Numerical values of sensory and motor nerve conduction studies of patients' lower extremities

| | | | | Sural Nerve | | | | Tibial Nerve | | | Peroneal Nerve | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | Sensory Nerve | | | | Motor Nerve | | | Motor Nerve | | |
| | | | | Right | | Left | | | | | | | |
| | n | Mean of age (SD) | Female (n) | SNAP amplitude mean (SD) (µV) | NCV mean(SD) (m/s) | SNAP amplitude mean(SD) (µV) | NCV mean(SD) (m/s) | DML mean(SD) (ms) | CMAP amplitude mean(SD) (mV) | NCV mean(SD) (m/s) | DML mean(SD) (ms) | CMAP amplitude mean(SD) (mV) | NCV mean(SD) (m/s) |
| Toxic | 11 | 64.1(9.9) | 4 | 0.4(0.95) | 48(5.6) | 0.37(1.06) | 44(0) | 4.03(0.72) | 6.1(3.4) | 44.7(5.7) | 4.43(0.74) | 2.1(1.7) | 43.9(3.4) |
| Idiopathic | 14 | 56.09(17.3) | 4 | 0.26(0.87) | 44(0) | 0.56(1.2) | 42(0) | 4(1.02) | 6.5(4.2) | 44.3(6.6) | 4.27(0.66) | 2.8(1.7) | 43.7(2.4) |
| Friedreich ataxia | 6 | 30.1(8.2) | 4 | 0(0) | Ø | 0(0) | Ø | 3.56 (0.82) | 8.9(2.8) | 45.8(4.7) | 4.5(1.01) | 2.7(2.1) | 44.4(4.2) |
| Sjögren's+ Sicca syndrome | 6 | 54.6(21.2) | 4 | 0(0) | Ø | 0(0) | Ø | 4.03(0.87) | 9.4(2.5) | 43.5(3.5) | 3.92(0.28) | 4.06(1.7) | 43.8(3.3) |
| CANVAS | 3 | 59.3(5.7) | 3 | 0.83(1.4) | 49(0) | 0.65(0.9) | 46(0) | 3.56(0.32) | 5.8(2.5) | 42.3(2.08) | 5.13(1.26) | 1.66(1.2) | 48.6(6.1) |
| Paraneo-plastic | 3 | 54.6(17.09) | 2 | 0(0) | Ø | 9(0) | 55(0) | 3.83(0.2) | 7.1(4.8) | 42.6(3.7) | 4.63(0.2) | 1.53(0.05) | 45.6(5.6) |
| HSAN II | 1 | 34 (0) | 0 | 0(0) | Ø | 0(0) | Ø | 3.9(0) | 4(0) | 50(0) | 4.4(0) | 2.4(0) | 41(0) |
| Celiac disease | 1 | 24(0) | 1 | 0(0) | Ø | 0(0) | Ø | 2.8(0) | 16(0) | 42(0) | 3.3(0) | 4.2(0) | 49(0) |
| Miller Fisher syndrome | 1 | 34(0) | 0 | 11.7(0) | 53(0) | 7.9(0) | 38(0) | 4(0) | 11.1(0) | 46(0) | 4.1(0) | 5.6(0) | 43(0) |
| Kennedy disease | 1 | 46(0) | 0 | 0(0) | Ø | 0(0) | Ø | 3.2(0) | 2.9(0) | 52(0) | 5(0) | 3.6(0) | 43(0) |
| FOSMN | 1 | 53(0) | 0 | 4.4(0) | 53(0) | Ø | Ø | 4.5(0) | 11.2(0) | 41(0) | 6.5(0) | 0.4(0) | 43(0) |
| All patients** | 48** | 54(18.7) | 25 | 0.54(1.79) | 55(0.62) | 0.76(2.02) | 46.8(6.06) | 3.97(0.94) | 7.3(3.9) | 44.4(5.1) | 4.44 (0.78) | 2.7(1.8) | 44.9(4.2) |

* n: Number; SD: Standard Deviation; SNAP: Sensory Nerve Action Potential; NCV: Nerve Conduction Velocity; DML: Distal Motor Latency; CMAP: Compound Muscle Action Potential; CANVAS: Cerebellar ataxia, Neuropathy and Vestibular Areflexia Syndrome; HSAN II: Hereditary Sensory and Autonomic Neuropathy Type II; FOSMN: Facial Onset Sensory and Motor Neuronopathy; Ø: Not performed
**Data of the remaining 8 patients who did not attend follow-up visits and therefore whose etiology could not be determined were not reported in the table.

**Table 3.** Data of SEP, Blink Reflex and HRV studies of the patients

| | n | Number of Tibial SEP | Number of Blink Reflex Study | Number of HRV |
|---|---|---|---|---|
| Toxic | 11 | Ø | 1(normal) | Ø |
| Idiopathic | 14 | 1(conduction impairment) | Ø | 1(autonomic dysfunction) |
| Friedreich ataxia | 6 | 2(conduction impairment) | Ø | Ø |
| Sjögren's+Sicca syndrome | 6 | 1(conduction impairment) | 1(normal)+1(unilateral R1 latency prolonged) | Ø |
| CANVAS | 3 | Ø | 1(bilateral R1 latency prolonged) | 1(autonomic dysfunction) |
| Paraneoplastic | 3 | Ø | Ø | Ø |
| HSAN II | 1 | Ø | Ø | 1(autonomic dysfunction) |
| Celiac disease | 1 | 1(conduction impairment) | Ø | Ø |
| Miller Fisher syndrome | 1 | Ø | 1(normal) | Ø |
| Kennedy disease | 1 | 1(conduction impairment) | Ø | Ø |
| FOSMN | 1 | Ø | 1(bilateral ipsi and contralateral R2 latency prolonged) | Ø |
| *All patients*** | 48** | 11 | 5 | 3 |

*n: Number; SEP: Somatosensory Evoked Potential; HRV: Heart Rate Variability; CANVAS: Cerebellar ataxia, Neuropathy and Vestibular Areflexia Syndrome; HSAN II: Hereditary Sensory and Autonomic Neuropathy Type II; FOSMN: Facial Onset Sensory and Motor Neuronopathy; Ø: Not performed
**Data of the remaining 8 patients who did not attend follow-up visits and therefore whose etiology could not be determined were not reported in the table.

**SEP/Blink Reflex/HRV Studies**

Table 3 shows the data of SEP, blink reflex and HRV studies of the patients.

Tibial SEP studies was applied to 11 patients. While conduction abnormality in both the central and peripheral sensory pathways was reported in 9 patients, conduction abnormality in the central sensory pathways of the posterior cord was reported in only 2 patients.

The results were normal in 3 of 6 patients who underwent blink reflex study, and the etiologies of these patients was MFS, alcoholism and Sjögren's-paraneoplastic syndrome coexistence. It was noted that R1 latencies were prolonged in one CANVAS and Sjögren's syndrome patient each, and prolonged R2 latencies were noted in FOSMN patient.

HRV was performed in 3 patients with CANVAS, HSAN II and idiopathic etiology and showed autonomic system disorder in all of them.

**Other Supportive Tests**

Spinal MRI, encompassing the cervical spine, was performed in 22 patients. Of these, 12 exhibited T2-weighted hyperintense lesions, either isolated to the cervical spine (Fig. 2a-b) or extending throughout the entire spinal dorsal column (Fig. 2c-d).

Epidermal nerve fiber density was not evaluated by skin biopsy in any patient. Sural nerve biopsy was performed in 4 patients, but as expected, no informative or discriminatory results were obtained.

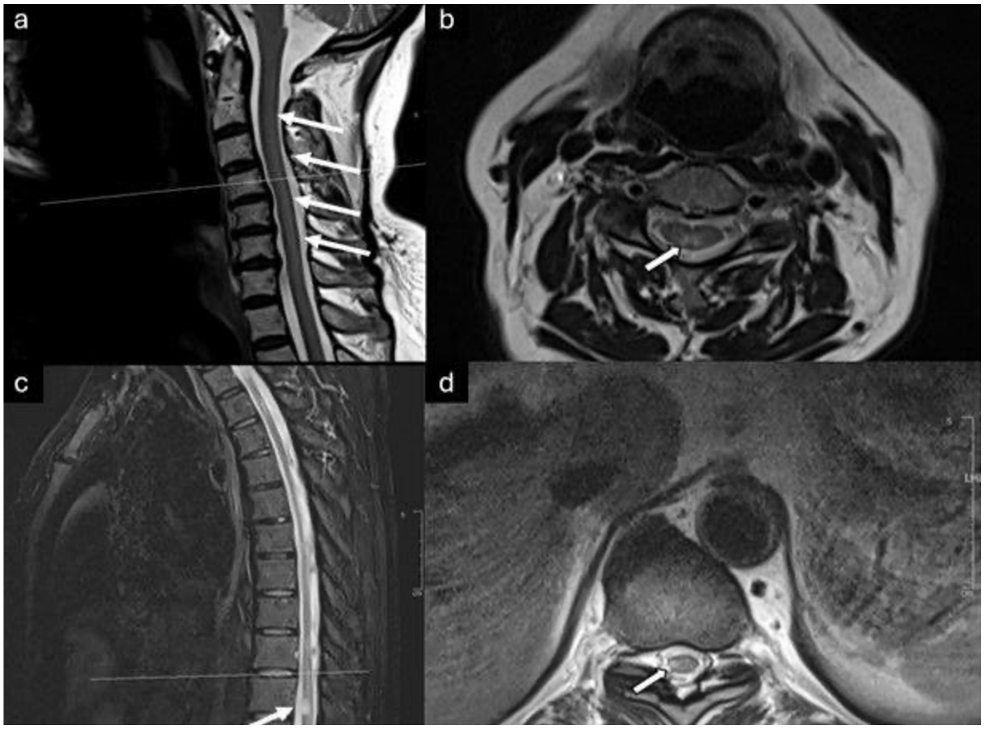


**Figure 2 (a-d).** Figure 2a-b are from a patient with Sjögren's syndrome, and figure 2c-d are from an anti-Hu seropositive patient (paraneoplastic syndrome). Sagittal and axial T2-weighted images show a longitudinal lesion *(arrows in a and b)* along the dorsal column in the cervical cord. Sagittal (c) and axial (d) T2-weighted spinal magnetic resonance images demonstrating a focal T2-hyperintense lesion within the thoracic spinal cord *(arrows in c and d)* in another patient.

## DISCUSSION

In this study, we reviewed electrophysiological tests, clinical features and supportive diagnostic tests of 56 patients electrophysiologically diagnosed with SG from a complementary approach. We confirmed that in the case of SG suspicion, comprehensive evaluation by electrophysiological methods and detailed examination is necessary because the definitive diagnosis is made by DRG biopsy, which is not routinely recommended due to the fact that it is an invasive procedure. In addition, sometimes SG can be diagnosed electrophysiologically after EDx without any prior clinical suspicion.

In our study, etiology could be determined 62.5% of all patients besides a considerable idiopathic preponderance (n=14; 25%) but not in agreement with literature. This rate was reported to be around 50% in the literature (11, 12).

Diagnosing and distinguishing SG from other relatively more common peripheral neuropathies is challenging. On the one hand, it is necessary to be aware of such situations that SG may appear several months after the completion of some chemotherapy treatments (coasting effect) (7); on the other hand, it is necessary to know that there are reversible underlying etiologies. Therefore, the distinction of SG is particularly required for the reversible causes. Due to diagnostic difficulties the ulnar sensory-motor amplitude ratio (5) and the proximally evoked soleus H-reflex (13) were also described as the neurophysiological methods for screening for SG. Based on the need for both sensitive and easy-to-use criteria, Asbury's criteria, causing hesitations in specificity and sensitivity evaluations (3), and PNS Euronet group criteria that were not reliable with other forms of SG except for paraneoplastic SG (3), were proposed. Recently, more comprehensive and highly accurate Camdessanché criteria (3) were suggested. Camdessanché criteria consist of a combination of three clinical and two neurophysiological parameters. The two neurophysiological criteria are 1) at least 1 sensory potential absent or amplitudes of 3 SNAPs <%30 of the lower limit of normal in the upper limbs, not explained by entrapment neuropathy and 2) less than two nerves with abnormal motor NCS in the lower limbs. Camdessanché criteria include MRI and DRG biopsy for diagnosis but do not mention skin biopsy. Currently, no method or criterion can distinguish SGs etiologically from each other.

In our patient group, we found that ulnar nerve DML was at the upper limit or prolonged in 27 patients (48.2%) and the lack of significance in the comparison of ulnar nerve DMLs of 23 patients diagnosed with pure SG and 9 SG patients diagnosed with motor axonal involvement. It may be possible that these findings could be explained by the relatively greater involvement of upper extremity nerves and mild motor fiber involvement in SG. As a matter of fact, Camdessanché and colleagues already found that the nerves in the upper extremities were significantly more compromised in SG and that limited electrophysiological motor nerve involvement is expected in SG (3). However, our interpretation may be excessive due to the sex and extremity length differences and the issue of unstandardized distance due to the retrospective study design. On the other hand, if a prospective study in SG patients is designed in the future in which the distances for ulnar and even radial distal motor latency measurements could be standardized, the question also arises whether the DMLs of these nerves would be at the upper limit/prolonged.

Besides 9 SG patients with mild motor axonal involvement findings, there were 19 patients with chronic diffuse or segmental root/anterior horn involvement. Moreover, needle EMG findings were detected in the lower extremities in all patients and in the upper extremities in 84.6% of the patients who underwent needle EMG procedure. Regarding the cause of motor fiber involvement, there are views that some patients may have anterior horn involvement similar to DRG (14) and/or peripheral nerves being affected due to a specific or nonspecific immune responses (15). Moreover, it is stated that motor fiber involvement may be more severe in paraneoplastic neuronopathies (11). However, only 1 of our 4 paraneoplastic SG patients had mild motor axonal involvement in our study. The diagnoses of 2 of our 3 patients with grade 2 UEMG findings were confirmed as CANVAS and Kennedy disease. The etiologies of 3 of our 7 patients with grade 2 LEMG findings were detected as toxic etiology, CANVAS and Kennedy disease. Our only patient with grade 3 needle EMG findings in the lower and upper extremities was FOSMN patient.

Involvement of the posterior column of the spinal cord can be more accurately demonstrated by MRI rather than SEP (2). Because SEPs can be obscured by low amplitude or absence of SNAPs (16), like in 9 of our 11 patients who underwent tibial SEP. This is a limitation of SEP in the diagnosis of SG. Autonomic ganglion involvement may also accompany with SG (17). Especially when specific etiologies are addressed, autonomic tests such as HRV and sympathetic skin response are helpful in proving autonomic involvement as in our 3 patients. Supportive electrophysiological tests such as SEP -despite its limitations-, blink reflex study and HRV can assist specialists in many clinics by suggesting and supporting the diagnosis of "*neuronopathy*", rather than making a hasty diagnosis of "*peripheral axonal neuropathy*" in certain patients. These test also help to narrow the differential diagnosis spectrum and facilitate rapid identification of the diagnosis and treatable etiologies. Perhaps, the more frequent use of these tests in clinics and the reporting of their findings will enable them to be included in the diagnostic criteria of SG as supportive tools in the future.

Sural nerve biopsy, which usually shows nonspecific loss of mostly large-diameter nerve fibers, may sometimes support the diagnosis of SG if the regeneration clusters are absent or the number of regeneration clusters is less compared to the number of lost fibers. However, this finding is not very helpful in the diagnosis of SG (11). Distinct skin biopsy pattern showing that intraepidermal nerve fiber density in the proximal thigh is lower than in neuropathies and controls, while in the distal leg it is at a value between neuropathies and controls is a valuable additional investigation tool (5, 18). Regrettably, epidermal nerve fiber density was not evaluated by skin biopsy in any of our patients.

This observational study has a number of limitations, starting with its own retrospective nature. The other main limitation is the lack of histopathological DRG examination of the patients who were acknowledged to have SG based on EDx tests.

As a result, in order to diagnose SG as early as possible without the need for invasive methods and to elucidate its etiology, a thorough electrophysiological screening should be performed with the help of other supportive diagnostic tools.

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