SENSORY GANGLIONOPATHY PRESENTING WITH SEVERE ATAXIC GAIT IN A MIDDLE-AGED FEMALE WITH REVIEW OF LITERATURE

K Hebishi, A Hebishi, R Donathan, M Lang

Introduction

Sensory Ganglionopathy (SG) is a rare condition characterized by selective destruction of dorsal root ganglia (DRG) neurons in the spinal cord and trigeminal ganglia. SG was first described in 1948 by Denny Brown (Denny-Brown, 1948). The sensory ganglia contain the cell bodies of the sensory nerves. They are located on both sides of the spinal cord and attached to the dorsal root of each spinal nerve. Their proximal projections are the sensory nerve roots that enter the spinal cord, and their peripheral projections are the sensory fibers of peripheral nerves. Those ganglia are susceptible to autoimmune targeting due to their fenestrated endothelial cells that form a permeable blood–nerve barrier (Amato & Ropper, 2020). DRG disease leads to clinical presentation of sensory loss including sensory ataxia (Camdessanche et al., 2009).

SG has a distinctive pattern from other peripheral neuropathies. In SG the damage is confined to sensory neuron causing only sensory loss. In peripheral neuropathy there is commonly axonal damage (e.g., in ischemic or diabetic neuropathy) which leads to symptoms starting distally and migrating proximally. In SG sensory features can start in any or all territories innervated by sensory nerves, particularly in the face, scalp, oral mucosa, trunk, and proximal limbs. A detailed comparison between SG and peripheral neuropathy is provided in Table 1.

Challenges of sensory ganglionopathy identification remain present despite availability of diagnostic criteria (table 1). In up to 50% of cases no identifiable cause can be found despite extensive workup (Amato & Ropper, 2020). The rarity of the condition and unfamiliarity of internists can delay diagnosis and treatment. Early Intervention can halt progression and prevent disability (Sheikh & Amato, 2010). Major distinctive difference from common causes of peripheral neuropathy are summarized in Table 2.

Case

Subjective:
- HPI: Middle aged female with no known personal family history presented to the neurology clinic with complaint of 1 year of progressive limb sensory deficits with numbness and tingling and subjective feeling of weakness. This has progressed to severe gait disturbance with inability to walk and frequent falls. Patient was treated for the past year for presumptive lumbar radiculopathy with no obvious explanation to upper extremity dysfunctions. She was also referred to psychiatry for a presumption of a functional neurological disorder and patient was started on Lexapro for anxiety. Over the course of her illness, she was referred also to pain management for local lumbar injection with minimal or no improvement in symptoms.
- ROS: Positive for occasional tension headaches, Persistent negatives: No Fever, cough, SOB, N, V or diarrhea.
- Medical history: Anxiety and lumbar radiculopathy. Negative for diabetes, hypertension, or peripheral vascular disease.
- Surgical history: none.
- Family history: no history of similar condition in her family.
- Past medical history: none.
- Allergies: none.
- Medications: Retuximab.
- A diagnosis of SNN is definite if dorsal root ganglia degeneration is pathologically demonstrated although dorsal root ganglia biopsy is not recommended.

Objective:
- □ Physical Exam: Positive for: decreased muscle tones all over, Loss of proprioception, Ataxia, decreased sensation to fine touch and vibration, severe truncal ataxia and Pseudohypertrophy (a search of position in space), Romberg’s sign, loss of biceps, triceps and Achilles’ tendon reflexes. Decreased quadriceps reflex

Management:
- Given severity of presentation patient was started on PLEX and showed progressive improvement over time. By the end of 6th session patient was able to walk with a walker. This was followed by a 3-week inpatient rehabilitation program. Patient was discharged home with ability to walk unassisted.

Sensory Ganglionopathy distinctive features

1. No motor loss.
2. does not start distally and migrate, can start in any area along the sensory tract.
3. Most common areas to start: Face, Scalp, Oral Mucosa, Trunk or proximal limbs.
4. SG has more focal loss of touch, vibration, proprioception and Pseudohypertrophy (a search of position in space), Romberg’s sign, loss of tendon reflexes without weakness.
5. Can rarely affect small fibers causing burning, tingling and stabbing pain.
6. Often associated with damage to the autonomic ganglia leading to dysautonomia (e.g., orthostatic hypotension, cardiac arrhythmia, gastriac dysmotility, and sweating disturbances).
7. Can affect spinocerebellar tract causing a specific form of ataxia that simulates cerebellar disorder but without dystonia and nystagmus.