A CASE OF PROGRESSIVE GENERALISED WEAKNESS WITH WASTING

Case presentation – Department of General Medicine

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1. PATIENT PROFILE

Name	Laxmi Barad
Age / Sex	65 years / Female
Religion	Hindu
Occupation	Farmer
Address	Chandipadar, Ganjam
Date of admission	08 / 08 / 26

2. CHIEF COMPLAINTS

Complaint	Duration
Weakness of both lower limbs	2 years
Weakness of left upper limb	2 years
Weakness of right upper limb	1 year
Slurring of speech	1 year
Difficulty in swallowing	3 months

3. HISTORY OF PRESENT ILLNESS

Patient was apparently alright 2 years back, when she developed weakness of the **left lower limb**, insidious in onset and gradually progressive. It manifested as tripping while walking with two episodes of fall, inability to grip the slippers and slippage of slippers while walking, of which she was aware. After 15 days she was walking with the help of a stick.

After 1 month she developed weakness of the **right lower limb**, insidious in onset and gradually progressive, such that she had to walk holding on to walls and was unable to climb stairs or get up from a squatting position without help. Since 3 months she was moving in the sitting position by dragging her lower limbs, and since 15 days she has been bed-ridden.

Simultaneously she had **twitching movements** in both upper and lower limbs.

She developed weakness of the **left hand**, such that she was unable to milk the cows; relatives noticed clawing of the left hand. Since 1 year she developed weakness of the **right hand** in the form of difficulty in mixing food and inability to open the cap of a bottle. Relatives noticed thinning of both lower limbs since 6 months.

Since 1 year she developed **difficulty in speech**, insidious in onset, with difficulty in initiating speech and straining while speaking; it progressed over 6 months such that relatives found her speech difficult to understand. Since 1 month she is unable to speak, though she understands words and responds to commands by gesturing. This is associated with inappropriate laughing and crying.

Since 3 months she has **difficulty in swallowing**, initially to solids and later to liquids, associated with drooling of saliva and not associated with nasal regurgitation.

Negative history

- No difficulty in breathing.
- No difficulty in rolling from side to side.
- No bowel or bladder involvement.
- No sharp shooting pain radiating to arm or back.
- No nasal intonation of voice.

4. PAST, FAMILY AND PERSONAL HISTORY

Past history

- Not a known case of hypertension, type 2 diabetes mellitus or tuberculosis.
- No contact history of tuberculosis.

Family history

- No significant family history.

Personal history

- Mixed Indian diet; good appetite; normal sleep pattern.
- No addictions.
- Married since 45 years, blessed with 5 children.

Menstrual history

- Attained menopause at 48 years of age.

5. SUMMARY OF HISTORY

A 65-year-old female presented with gradual onset, slowly progressive, **asymmetrical** distal and proximal weakness of both lower limbs and distal weakness of both upper limbs, with atrophy of upper and lower limbs, dysarthria and dysphagia, **without** sensory symptoms and **without** bowel or bladder involvement.

6. GENERAL EXAMINATION

Patient is conscious and cooperative.

Pulse	85/min, regular in rhythm, normal volume and character; no radio-radial or radio-femoral delay; all peripheral pulses felt bilaterally and equally
Respiratory rate	18/min, regular, thoraco-abdominal
Blood pressure	100/60 mmHg, right arm, supine
Temperature	97.8 °F

- No pallor, icterus, cyanosis, clubbing, oedema or lymphadenopathy.
- No thyromegaly. No neurocutaneous markers.

7. CENTRAL NERVOUS SYSTEM

A. Higher mental functions

- Conscious, oriented to time, place and person.
- Right handed. Memory intact.
- Speech – **spastic dysarthria**.
- No hallucinations or delusions.

B. Cranial nerve examination

Table 1. Cranial nerves I–V

Cranial nerve	Right	Left
I – Olfactory	Normal	Normal
II – Optic		
Visual acuity	Normal	Normal
Colour vision	Normal	Normal
Field of vision	Normal	Normal
III, IV, VI		
Extraocular movements	Normal	Normal
Pupils	Normal	Normal
Light reflex	Normal	Normal
V – Trigeminal		
Sensation over face	Normal	Normal
Motor	Normal	Normal
Corneal reflex	Present	Present
Jaw jerk	Present	—

Table 2. Cranial nerves VII–XII

Cranial nerve	Right	Left
VII – Facial		
Forehead wrinkling	Present	Present
Angle of mouth	Not deviated	Not deviated
Taste sensation	Intact	Intact

Cranial nerve	Right	Left
VIII – Vestibulocochlear Rinne test Weber test	 AC > BC Not lateralized	 AC > BC Not lateralized
IX, X – Glossopharyngeal, Vagus Uvula Palatal movements Gag reflex	 Central Normal Present	 Central Normal Present
XI – Spinal accessory Trapezius Sternocleidomastoid	 Normal Normal	 Normal Normal
XII – Hypoglossal Tongue	 **Fasciculations and atrophy; no deviation of tongue**	 —

C. Motor system – bulk

Segment	Right	Left
Arm	21 cm	21 cm
Forearm	18 cm	18 cm
Thigh	29 cm	30 cm
Leg	18 cm	18 cm

Atrophy of thenar muscles of both hands and of both lower limbs; fasciculations present.

D. Tone

• Upper limbs – normal tone.

• Lower limbs – hypotonia.

E. Power – upper limb (MRC grade)

Joint	Movement	Rt	Lt
Shoulder	Adduction	5	5
	Abduction	5	5
	Flexion	5	5
	Extension	5	5
Elbow	Flexion	5	5
	Extension	5	5
Wrist	Flexion	3	3
	Extension	3	3
Hand grip		Normal	Weak

Small muscles of the hand

Muscle	Right	Left
Abductor pollicis longus	Weak	Weak
Abductor pollicis brevis	Weak	Weak
Extensor pollicis longus	Weak	Weak
Extensor pollicis brevis	Weak	Weak
Opponens pollicis	Weak	Weak
Flexor pollicis longus	Weak	Weak
Adductor pollicis	Weak	Weak
Palmar interossei	Weak	Weak
Dorsal interossei	Weak	Weak
Lumbricals	Weak	Weak
Flexor digitorum sublimis	Weak	Weak
Flexor digitorum profundus	Weak	Weak

F. Power – lower limb (MRC grade)

Joint	Movement	Rt	Lt
Hip	Flexion	1	1
	Extension	1	1
	Abduction	1	1
	Adduction	1	1
Knee	Flexion	2	2
	Extension	1	1
Ankle	Dorsiflexion	1	1
	Plantar flexion	1	1
	EDL	1	1
	EHL	1	1

G. Reflexes

Deep tendon reflex	Right	Left
Biceps	2+	2+
Triceps	2+	2+
Supinator	2+	2+
Knee	3+	3+
Ankle	2+	2+

Clonus – absent.

• Superficial reflexes: abdominal reflex present.

• Plantar response: right – no response; left – no response.

H. Cerebellar signs and gait

• Cerebellar signs – absent.

• Gait – could not be tested.

I. Sensory examination

• All primary modalities – touch, pain, temperature, proprioception and vibration – intact.

• All cortical sensations intact.

J. Autonomic nervous system

• No resting tachycardia.

• Bowel and bladder sensation and control normal.

• No postural hypotension.

K. Skull and spine

• No deformity.

8. OTHER SYSTEMS

Respiratory system

• Chest movements equal bilaterally.

• Bilateral vesicular breath sounds heard equally over all lung areas; no added sounds.

Cardiovascular system

• Apex beat in left 5th intercostal space, ½ inch medial to the mid-clavicular line.

• First and second heart sounds normal; no murmur.

Gastrointestinal system

• Abdomen soft, non-tender; no organomegaly.

9. SUMMARY OF THE CASE

A 65-year-old female presented with gradual onset, slowly progressive weakness of both upper and lower limbs, **distal more marked than proximal**, with thinning of both upper and lower limbs, dysarthria, tongue atrophy and fasciculations, without sensory, cerebellar, bowel or bladder symptoms.

On examination she was found to have dysarthria, **pseudobulbar palsy** of the 9th and 10th cranial nerves and **bulbar palsy** of the 12th cranial nerve; signs of a **lower motor neuron** lesion in the upper limbs as evidenced by wasting of muscles, fasciculations and hypotonia, and signs of a lower motor neuron lesion in the lower limbs as evidenced by hypotonia and wasting, without any sensory, bowel, bladder or cerebellar involvement.

10. ANATOMICAL LOCALISATION

Structure involved	Function lost
1. Anterior horn cells	Fasciculations; atrophy of small muscles of the hand
2. Bilateral corticobulbar fibres to 9th and 10th cranial nerves	Spastic dysarthria
3. Nucleus of 11th cranial nerve	Weakness and atrophy of sternocleidomastoid and trapezius
4. Nucleus of 12th cranial nerve	Tongue atrophy and fasciculations
5. Corticospinal tract	Exaggerated deep tendon reflexes

Note: motor nuclei of the 9th and 10th cranial nerves – weak pharyngeal muscles.

11. PROVISIONAL DIAGNOSIS

Motor neuron disease – amyotrophic lateral sclerosis (ALS).

12. DIFFERENTIAL DIAGNOSIS

• Cervical myelopathy.

• Paraneoplastic motor neuronopathy.

• Cranio-vertebral junction anomaly.

Table 3. Points for and against each differential

Differential	Points in favour	Points against
Cervical myelopathy	Progressive weakness; asymmetrical; distal to proximal weakness	No bowel or bladder involvement; no sensory involvement; no root pain
Paraneoplastic	Old age	No respiratory symptoms; no significant weight loss; expected rapid progression absent
CV junction anomaly	Progressive weakness; cranial nerve 9–12 involvement; wasting of small muscles of hand and upper limb	No short neck; no low hairline; cerebellar signs absent