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REVIEW ARTICLE

PRIMARY LATERAL SCLEROSIS: A RARE CASE REPORT

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ABSTRACT

Primary Lateral Sclerosis (PLS) is an uncommon motor neuron disorder. Primary lateral sclerosis (PLS) is characterized by insidious onset of progressive upper motor neuron dysfunction in the absence of clinical signs of lower motor neuron involvement . The course of the disease is insidious and progressive, usually starting with the lower extremities, weakness may progress to affect the arms and the muscles at the base of the brain (bulbar muscles). As a rare disease, diagnosis is exclusionary. We are reporting a case of 46 year old female presented with lower limb weakness since 3 years which progressed to upper limb weakness and spastic dysarthria.

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INTRODUCTION

Primary lateral sclerosis (PLS) is a disorder of progressive upper motor neuron dysfunction, in the absence of clinical signs of lower motor neuron involvement or family history suggestive of hereditary spastic paraplegia. PLS is a diagnosis of exclusion. PLS exists on a spectrum of sporadic motor neuron disorders, including progressive muscular atrophy (lower motor neuron only), and amyotrophic lateral sclerosis (mixed upper and lower motor neuron involvement). PLS is a rare disorder[4], representing approximately 1–4% of all patients with motor neuron disease[1-3]. PLS typically presents in patients in their early 50s.

The rate of progression can be exceedingly slow, often progressing over many years to the point where the patient manifests a robotic gait, debilitating generalized spasticity,emotional lability and prominent pseudobulbar palsy. Muscle atrophy, if it occurs at all, is a very late feature. No clinically detectable sensory changes occur[5].
PLS is characterized by progressive weakness and stiffness in voluntary muscles, typically starting in the lower extremity. The disease may then progress to the trunk, followed by the upper extremity, and lastly to the corticobulbar tract, typically causing a pseudobulbar affect (emotional lability). In addition, patients may also experience dysarthria (difficulty speaking) and dysphagia (difficulty swallowing).

Diagnostic criteria By Pringles and Cols

Clinical hallmarks of PLS include[6]:

- Insidious onset of stiffness, clumsiness or mild weakness; or dysarthria, dysphagia, and emotional lability
- Symptoms begin most commonly in the legs, but can begin in the bulbar region or multiple areas of the body
- Signs include spasticity, hyperreflexia, and upper motor neuron pattern weakness
- The absence of diffuse fasciculations or muscle wasting, or sensory symptoms or signs
- PLS is progressive, spreading from side to side and from region to region
- Urinary urgency or frequency may be reported

Pringle criteria symptoms had to be present ≥3 years; in the Singer criteria ≥4 years; and in our ongoing COSMOS study in PLS patients had to have symptoms ≥5 years. But common features include the clinical presence of:

- Upper motor dysfunction on exam – spasticity, pathological reflexes, and upper motor neuron pattern of weakness
- Presentation most commonly in the legs, but can be in the bulbar region, or mixed limb and bulbar

- Slow progression of symptoms (≥4 years) with an age of onset ≥ 20 years in absence of:
- Marked fasciculations or muscle atrophy
- Sensory signs on exam
- Family history of similar disorder
- In addition laboratory or diagnostic studies must be negative for an alternative explanation for the symptoms. Additional normal studies supportive of PLS include:
- B12, copper, HTLV1/2, HIV testing, paraneoplastic workup
- MRI of brain and spine
- CSF evaluation
- EMG (normal, or minimal denervation that does not fulfill El Escorial criteria)

Diagnostic criteria proposed by Pringle and Cols.

- Clinical:

1.- Insidious onset of spastic paresis, usually starting in lower extremities, but occasionally in bulbar system or upper extremities.
2.- Start in adulthood, usually in the fifth decade of life or after.
3.- Absence of family history.
4.- Gradually progressive course.
5.- Duration greater than or equal to three years.
6.- Clinical findings generally limited to dysfunction of the corticospinal system.
7.- Symmetrical distribution, with final development of severe spinobulbar spastic paresis.

- Laboratory (for differential diagnosis).

1.- Blood chemistry in normal serum including vitamin B12 levels.
2.- Negative serology for syphilis and in endemic areas for Lyme disease and HTLV-1 virus.
3.- Normal CSF parameters including absence of oligoclonal bands.
4.- In most patients lack potential straining in GMS, occasional fibrillation and increased insertional activity in few muscles. Absence of compressive lesions of the cervical marrow or foramen magno in images of MRI-

- In addition to primary lateral sclerosis.

1.-Preserved bladder function.
2.- Absence or very prolonged latency in cortical evoked motor responses in the presence of normal peripheral evoked stimuli combined with potentials of muscle action.
3.-Focal atrophy of the pre-central rotation in MRI.
4.- Decreased glucose consumption in the pericentral region in PET.

Imaging findings in PLS include the following[7,8]:

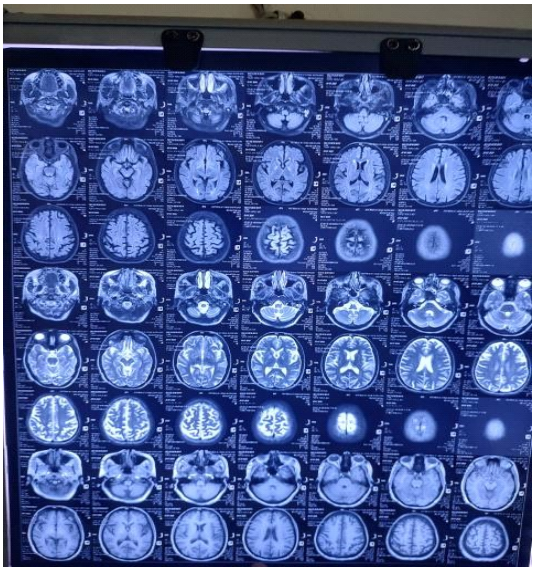
- Diagnostically MRI in PLS should be without structural abnormalities, with the exception of atrophy of the precentral gyrus
- MRI T2 imaging hyperintensity can be seen in the corticospinal tracts, which corresponds to decreased fractional anisotropy and increased mean diffusivity on DTI
- Metabolic imaging shows decreased function in the precentral gyrus (MRS, PET)

No specific pharmacotherapy is available. The mainstay of therapy in PLS remains supportive and is limited to multidisciplinary interventions to improve mobility, reduce muscle tone and facilitate activities of daily living[9]. However, antispasticity drugs such as the GABA-B agonist, baclofen, and the central α2-agonist, tizanidine, may be tried for symptomatic treatment. Severe spasticity sometimes requires the insertion of an intrathecal baclofen pump. Tricyclic antidepressants, selective serotonin reuptake inhibitors, or dextromethorphan/quinidine may control pseudobulbar affect lability. The prognosis is significantly better than for MND/ALS: one series had a median disease duration of 19 years and another series exhibited a range of survival from 72 to 491 months[10,11.]

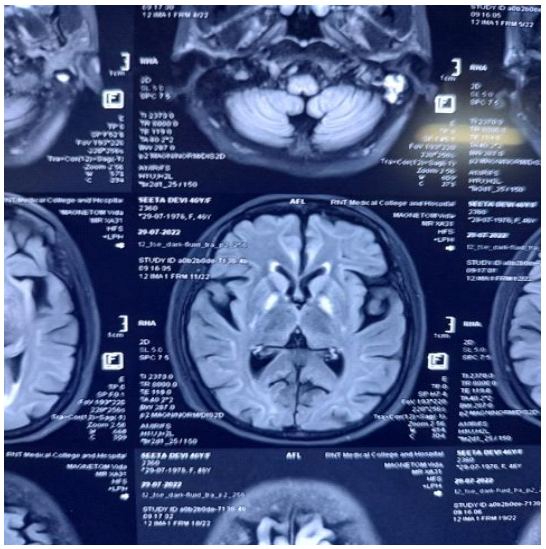
CASE REPORT

A 46 year old female with no comorbidity and no family history was admitted to our hospital with complaints of both lower limb weakness since 3 years and both upper limb weakness since 2 months. The patient was asymptomatic 3 years back when the patient experienced weakness in both the lower limbs insidious in onset gradually progressed such that the patient has difficulty standing then to both upper limbs.

On examination: mental functions were normal
Spastic dysarthria present
Cranial Nerves – Normal
Motor- bulk normal ; spasticity in all 4 limbs;
Power 4/5 in all 4 limbs
Hoffman sign - positive
Finger flexion sign - positive
Wartenberg's sign- positive
Planter-B/L extensor
Sensory examination: normal
Hb- 9.2g/dl ,Platelet count- 1,72,000/mm3,SGOT/SGPT-28/21,ALP-96,,TB/CB-1.3/0.328,Total Protein/Albumin- 6/3.42
Vitamin B12- 432 pg/ml
HIV-NR
VDRL-NR, HBsAg & Anti HCV – Negative
CSF study: 4 cells, normal sugar and protein, negative for oligoclonal bands
MRI Brain: T2W& FLAIR – Hyperintense signals seen in B/L Capsuloganglionic region & Precentral Gyrus Atrophy.
MRI spinal cord: no abnormality.
The patient was treated with Baclofen, regular physiotherapy and advised regular follow up in neurology.



MRI Brain Showing Precentral Gyrus Atrophy



MRI Brain T2WI Showing showing Hyperintensity in B/L Corticospinal tract

CONCLUSION

PLS is a rare motor neuron disease with diagnosis of exclusion typically presenting as symmetrical spastic paraparesis progressing slowly to quadriparesis without sensory involvement with variable prognosis. Treatment is symptomatic, supportive care and physiotherapy.

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