Case study
Amyotrophic lateral sclerosis with frontal dementia - A case study
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Abstract
Amyotrophic lateral sclerosis (ALS) or Lou Gehrig’s disease is the most common form of motor neuron disease characterized by the gradual degeneration and death of motor neurons. A patient is described in whom a profound and rapidly progressive dementia occurred in association with clinical features of Amyotrophic lateral sclerosis. We present here the case of 60 year old women with motor neuron disease, hypothyroidism, depression, osteoporosis and frontotemporal dementia commonly searched for disorders during ALS or motor neuron disease. Our intention is to report rare case like Amyotrophic lateral sclerosis with the review of literature.

Keywords: Amyotrophic lateral sclerosis, Electromyography, Dementia, Escorial criteria, CMAPs

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1. Introduction
Amyotrophic lateral sclerosis/Lou Gehrig’s/Charcot’s disease is a progressive and neurodegenerative disease involving both lower and upper motor neurons [1,2,3,4,5] typically following a relentless path towards death [1]. The wide spectrum of typical presentations can frequently lead to expensive work up and undue delay or failure in diagnosis of ALS in substantial number of patients [6,7,8]. This case was diagnosed based on (the revised El Escorial criteria) [9]. This is used to express in each patient the extent of disease involvement at the time of the examination. There is a need for the development treatment options that can aid in early diagnosis of ALS.

Case report
A 60-year old women housewife with 10 years of schooling presented with affective symptoms of memory, speech and swallowing disturbances for the past one year. She also showed swallowing difficulty more for solids, for liquids at times there is nasal regurgitation and choke episodes that had started six months earlier. In the previous six months, she had had a gait difficulty and walking difficulty with falls- a severe worsening of her behavioural disorder and of her language disturbance. The weakness in right upper limb since 5 months with subsequent progression to left lower limb. She also had a previous history of suicidal ideations and attempted suicide 20 years back. She was irregular on prescription since 5 years adherent to the prescription since 1 year. She was on prescription of fluoxetine 20 mg, Divalproex sodium 500 mg OD, Gabapentin 300mg and tablet olanzapine 10 mg for several months prior to her illness. In the patient, there was a previous psychiatric history. There had been no occupational exposure to chemicals, heavy metals, or previous family history of dementia. She had an accidental fall resulted in an injury to right wrist, hip and back and was evaluated only for the wrist. She wasn’t
evaluated for right hip pain and was lumping after one month she met with an accident and had injury to right shoulder and hip from then onwards she had faced difficulty in holding objects with right hand and wasty in right upper limb distally. She noted difficulty in lifting the right lower limb, foot drop and unable to wear slippers and falling forward even for small obstacles like mat, merely falling to right side. After the fall, she had a history of fasciculation’s in right upper limb, lower limb and back.

Findings and discussion

Amyotrophic lateral sclerosis is a motor neuron disease, which is performed by ENMG neurological examination. Nerve conduction studies and needle electromyography (EMG) are useful for confirming the diagnosis of ALS and for excluding peripheral conditions that resemble ALS. The present case showed that compound muscle action potential (CMAPs) could not be elicited from right posterior tibia and common peroneal nerves, CMAPs amplitudes are reduced and other parameters are within the normal limits in the left posterior tibia and common peroneal nerves. SNAP (sensory nerve action potential) could not be elicited from right sural nerve (due to oedema). On needle electromyography of right FDI, muscle showed spontaneous activity in the form of Fibrillations +2, positive sharp waves with very few recruit able MUP’s. Right EDC showed spontaneous activity in the form of fibrillations +2 with very few recruit able MUP’s. Right biceps muscle showed spontaneous activity in the form of fibrillations+3, fasciculation’s with MUPs of 4–6 mV amplitude, 10-12 ms with reduced recruitment. Tongue muscle showed spontaneous activity in the form of Fasiculations+1 proving of preganglionic neurogenic lesion with an evidence of denervation and renervation.

Fasciculation’s were noted in right upper limb and lower limb. Strength testing results using Medical Research Council grades were as follows: diminished grip strength (4/5) of the right hand also noted normal strengthening was 5/5. The snout reflex was present, finger flexion /pectoralis showed brisk, plannar group and glabbelar reflex, she had increased deep tendon reflexes. A mild gait imbalance was observed. Her sensory examination observed the decrease in pain and touch sensation in right lower limb below the knee.

Neuropsychological examination assessment showed no aggressive behaviour. Her spontaneous speech was hard to evaluate because of her spastic dysarthria on the visual naming scale she scored average. She was severely apraxic for right upper limb.

Table: Tabular representation of ALS with their condition

<table>
<thead>
<tr>
<th>Motor nerves</th>
<th>Lat (ms)</th>
<th>AMP (%)</th>
<th>FM (ms0)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left median Wrist-APB ELBOW-WRIST</td>
<td>3.5</td>
<td>-1</td>
<td>20.3</td>
</tr>
<tr>
<td>Left ulnar WRIST-ADM EOBOW–WRIST</td>
<td>2.2</td>
<td>-3</td>
<td>22.8</td>
</tr>
<tr>
<td>Right Tibia ANKLE-AHL KNEE-ANKLE</td>
<td>4.2</td>
<td>3</td>
<td>46.8</td>
</tr>
<tr>
<td>Left tibia Ankle–AHL Kne- ankle</td>
<td>5.6</td>
<td>-53</td>
<td>45.2</td>
</tr>
<tr>
<td>Right peroneal Ankle-EDB Fibula – ankle</td>
<td>3.8</td>
<td>-33</td>
<td>-</td>
</tr>
<tr>
<td>Left peroneal Ankle- EDB Fibula-ANKLE</td>
<td>4.5</td>
<td>-24</td>
<td>+42.0</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sensory nerves</th>
<th>Lat (ms)</th>
<th>AMP (mv)</th>
<th>CV (m/s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left medium DIGIT NO 2- WRIST</td>
<td>2.2</td>
<td>33</td>
<td>59.1</td>
</tr>
<tr>
<td>Left ulnar DIGIT NO 5- WRIST</td>
<td>1.67</td>
<td>29</td>
<td>65.9</td>
</tr>
<tr>
<td>Right sural Stim1-rec 1</td>
<td>1.96</td>
<td>14</td>
<td>51</td>
</tr>
<tr>
<td>Left sural Stim2-rec 2</td>
<td>2.2</td>
<td>20</td>
<td>54.5</td>
</tr>
</tbody>
</table>

Hallmark findings in the electro diagnosis of ALS are normal sensory nerve conduction studies and abnormal motor nerve conduction studies with
reduced motor compound muscle action potentials.

Radiographic findings of LS spine found to be wedge compression at D12 vertebral body, bone density is reduced, mild reduction in L3/L4, L4/L5 disc spaces. Osteoporosis of bones

• Right hip joint-osteoarthritis of right hip
• Right shoulder-bone density is reduced, decrease in acromion -c joint space

MRI of spine screening of whole spine shows C4 to C7 mild marginal osteophytes with desiccated discs. Extensive laboratory findings in these women did not reveal a cause for her disease. A complete blood count and basic metabolic profile were all within normal limits as were studies of vitamin B12. Recently she was diagnosed with hypothyroidism on medication thyroxin sodium 75 mg and Vitamin D total 21ng/mL showing insufficiency: 10-30ng/mL.

Cranial nerve examination revealed fasciculation’s of tongue with mild atrophy. Examination of extremities revealed muscle wasting with 2 to -4/5 asymmetric muscle power, power being less on the right side with prominence of proximal weakness. After reviewing old medical records, MRI and radiographic scans and considering the presence of both upper and lower motor neurons considered as Amyotrophic lateral sclerosis with frontotemporal dementia. ALS manifesting as upper motor neuron disease with bulbar signs and symptoms as the diagnosis. Even though the diagnosis was established from the clinical picture analyzed serum levels of vit B12, Folate, TSH, Vit D and heavy metals was also performed to confirm.

Conclusion

Amyotrophic lateral sclerosis is a rapidly progressive, fatal neurodegenerative disorder for which there is no effective treatment. This neurological study was carried by electromyography and a drug like riluzole is used to treat amyotrophic lateral sclerosis so thereby prudent survival rate can be maintained. Through our studies we addressed atypical presentation of ALS with frontal dementia. Patient counseling is necessary to improve the medication adherence.

Reference