Brugada Syndrome and Other Cardiac Issues in Amyotrophic Lateral Sclerosis

Rohit Gummi[1], Raghav Govindarajan MD[2]

[1] Medical Student, University of Missouri School of Medicine at Columbia, MO, 65201

[2] Department of Neurology, University of Missouri Hospital, MO, 65201

INTRODUCTION

Amyotrophic Lateral Sclerosis(ALS):

- ALS is a progressive motor neuron disease that results in the death of both lower and upper motor neurons.
- Weakness caused by denervation results in progressive atrophy of muscles and fasciculations. Onset can involve bulbar muscles, upper limb muscles, or lower limb muscles and progress to the others.
- Over the course of the disease, involvement of the muscles of respiration usually leads to death. Respiratory involvement is the most common cause of death.
- Previous studies have found links between ALS and decreased sympathetic activity which can lead to cardiac abnormalities and sudden death.[3]
- Here, we present 3 cases of cardiac manifestations in patients diagnosed with ALS, including the first reported case of Brugada syndrome in an ALS patient.

Brugada Syndrome:

- Characterized by particular ST-segment elevations in leads V_1 to V_2 in the presence or absence of sodium channel blockers.
- SCN5A gene, which codes for the α-subunit of cardiac sodium channels, has been liked to Brugada.
- Clinical Manifestations: Syncope and cardiac arrest at rest. This can result in sudden nocturnal death.

	Kennedy Cohorts[4]	ALS Case
Age at onset	43.4 ± 11.0	48
Presentation	Syncope along with muscle weakness of limbs and dysphagia.	Progressive right upper limb weakness, and twitching all over body.
Medical History	Most common included hypertension, hypercholesterolemia, and hypertriglyceridemia	Vitamin D deficiency, Anemia, papillary carcinoma of thyroid
ECG Abnormalities	35% had Type 1 41% had Type 2 23% had Type 3	Type 2 abnormalities
Electrolytes	140.7 ± 2.3 mEq/L Sodium 4.3 ± 0.5 mEq/L Potassium	142 mEq/L Sodium 4.3 mEq/L Potassium

OBJECTIVES

1. **Define cardiac abnormalities seen among patients diagnosed with Amyotrophic Lateral Sclerosis**
2. **Describe the first case of Brugada syndrome in an ALS patient**

CASES

Brugada Syndrome

- A 45 year old woman presented with progressive right upper limb weakness and fasciculations.
- She was diagnosed with upper limb onset ALS after extensive work including EMG which showed denervation in three body regions.
- She was wheelchair bound and had developed bulbar symptoms when she presented with shortness of breath.
- Her FVC measured a month ago in the clinic was 50% predicted.
- To further evaluate the sudden onset shortness of breath, CT chest to rule out pneumonia and a CT angiogram to rule out pulmonary embolism were done. These were normal.
- An EKG obtained in the ER showed J point elevation with saddle shaped ST segment elevation predominantly in V1 and V2 consistent with Brugada syndrome type 2 (see arrows in Figure 2).
- Extensive work-up for reversible causes was non diagnostic.

Example Brugada EKG

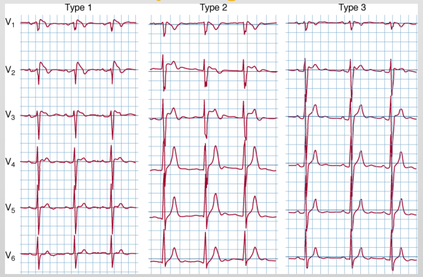

Figure 1: Characteristic EKG findings for type 1,2, and 3 Brugada Syndrome

Case EKG

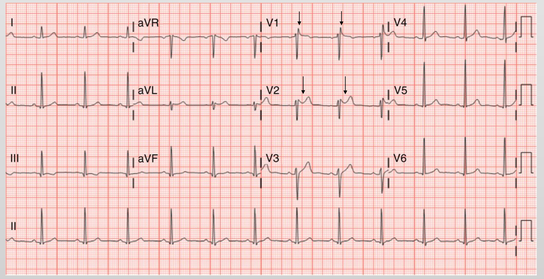

Figure 2: ALS patient's EKG showing Type 2 Brugada. Saddleback appearance can be seen in the precordial leads. This is characterized by a ≥2 mm ST elevation followed by a trough ≥1mm and either a positive or biphasic T-wave.

Dilated Cardiomyopathy

- A 70-year-old man presented with lower limb weakness and was wheelchair bound before he was diagnosed with ALS
- Three months later he presented with sudden onset shortness of breath.
- On evaluation, a CT of the chest showed pulmonary edema, and a CT angiogram did not show pulmonary embolus.
- An echocardiogram was performed and revealed an ejection fraction of 20% with dilated left ventricular cavity.
- Extensive workup for reversible causes was nondiagnostic.

Atrial Fibrillation

- A 62-year-old woman presented with slurred speech and difficulty in swallowing and was a diagnosed with ALS.
- Two months after diagnosis, she presented with sudden onset shortness of breath.
- On Evaluation, CT/angiogram of the chest was normal.
- An EKG revealed atrial fibrillation with rapid ventricular rate. She was treated with IV sotalol and placed on digoxin treatment.

CONCLUSION

- **Respiratory complications are most commonly seen with ALS**
- **Cardiac issues, although uncommon, could be a part of the clinical manifestation of ALS.**

DISCUSSION

- Patients with ALS often develop shortness of breath which can often be attributed to weakening of respiratory muscles. However, in some cases, it is a result of cardiac issues.
- Here, we presented 3 cases of ALS patients with cardiac issues. This supplements other cases of sudden cardiac deaths with ALS patients and introduces the first reported case of Brugada with ALS.
- Brugada syndrome has been seen in other instances of motor neuron disease such as Kennedy's Disease. Cardiac abnormalities such as Brugada might be a manifestation of motor neuron diseases themselves.
- Physicians should be aware that cardiac abnormalities can present with ALS and lead to death if not anticipated.

REFERENCES:

www.ALS-Brugada.xyz