

CLINICAL VIGNETTE

Arrhythmias and ECG Alterations in a Neuromuscular Disease Patient

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Abstract

Amyotrophic lateral sclerosis is a complex disorder with neurologic and cardiovascular sequelae. Evidence demonstrates that neuromuscular and autonomic imbalance contributes to ALS's cardiovascular effects. There exists a significant prevalence of abnormal ECG findings and arrhythmias in ALS. Physicians should recognize how ALS affects the cardiovascular system to allow for informed evaluation and management.

Case

A 77-year-old male with amyotrophic lateral sclerosis (ALS) complicated by ventilator-dependent respiratory failure, hypertension, and diabetes-mellitus was brought in to Emergency Department by paramedics for respiratory distress, lethargy, and tachycardia. Upon arrival of EMT to his home, he was unresponsive with a Glasgow Coma Scale (GCS) of 3 and was reported to have a heart rate of 164 bpm, with a reported "wide-complex tachycardia" on cardiac monitor. In the field, he received synchronized cardioversions up to 200J without a change in status.

On arrival to the ED, he was diaphoretic, tachycardic (158 bpm), hypertensive (163/101), febrile (40°C), and in respiratory distress (O₂-saturation 87%). ECG demonstrated a wide-complex tachycardia with a rate of 156 bpm. Administration of IV bolus 150 mg amiodarone and magnesium, followed by an amiodarone drip, improved his HR from 156 to 120 bpm. An old ECG was obtained, which demonstrated an existing RBBB and apparent "Q-waves" in leads II, III, aVF, and a QTc of 600ms. Cardiac electrophysiology (EP) was consulted for his wide-complex tachycardia and subsequent management.

Management

EP classified his initial SVT with aberrancy at a rate near 150 as typical atrial flutter with 2:1 conduction (Figure 1). Subsequent evaluation revealed urosepsis and pneumonia, and he was admitted to the MICU. Echocardiogram showed mild concentric left ventricular hypertrophy with an ejection fraction of 65% and normal wall motion. In addition to his atrial flutter, he demonstrated several other rhythms during hospitalization. On hospital day two, he had intermittent episodes of atrial fibrillation between sinus rhythm. By hospital day three, he

converted to predominantly sinus rhythm, but also demonstrated periods of isorhythmic dissociation with varying accelerated junctional and ventricular rhythms (Figures 2-4).

In the MICU, he transitioned from IV to PO amiodarone, with no further episodes of RVR during his hospitalization. Because his acute infections likely provoked his SVT and he eventually remained without further episodes, amiodarone was discontinued. Considering his ALS and to avoid precipitating heart block, no additional anti-arrhythmic medications were started. Ischemic evaluation was also deferred per patient request.

Discussion

The presence of multiple competing rhythms raised the question of what is known about the role of ALS in arrhythmogenesis. This case also helped clarify cardiovascular issues and rhythms typically found in ALS patients.

ALS is a progressive neurodegenerative disease characterized by the degeneration of both upper and lower motor neurons. It can also involve non-motor neural networks, including arrhythmogenic and autonomic disturbances on cardiac conduction.^{1,2} Due to the relatively small number of patients, as well as varied presentations, there are no current established guidelines on cardiac management in the ALS population.

Multiple studies have found impaired autonomic reflexes in ALS patients, with early sympathetic predominance and vagal withdrawal, followed by late-stage sympathetic denervation.¹⁻⁴ The etiology of these autonomic disturbances is multifactorial, including the effects of hypoxia from respiratory failure and neuropathological mechanisms resulting in autonomic nerve degeneration.² Postmortem biopsies found reduced numbers of neurons in the intermediolateral nucleus (IML) that give rise to the sympathetic nervous system. Decreased IML neurons are correlated with increased cardiac QTc dispersion, a marker of reduced sympathetic activity.³ ALS patients have arrhythmias and ECG changes ranging from ST-T-wave changes to complete heart block. Sudden cardiac death has caused death in patients with ALS and others with neuromuscular disorders, particularly those with right or left bundle branch block, bifascicular block, QT-prolongation, and ventricular

tachycardia.^{3,5} Diverse manifestations suggests complex autonomic neurodegeneration in ALS and impact on cardiac conduction. We review the arrhythmias seen in our patient.

Isorhythmic Dissociation

Our patient experienced episodes of isorhythmic dissociation with both sinus rhythm and sinus bradycardia with accelerated junctional rhythms. Isorhythmic dissociation is a form of AV-dissociation where the intrinsic sinus rate is approximately equal to another intrinsic cardiac rhythm (frequently a junctional rhythm of 50-60 bpm). This results in both the sinus and junctional pacemakers reaching each other in their respective refractory periods, usually with the presence of some antegrade or retrograde AV block. On ECG, the P waves and QRS complexes appear related, with P waves characteristically moving closer to, or even buried within, the QRS complex. Unlike complete heart block, fusion or capture beats can be seen. Isorhythmic dissociation is a benign arrhythmia that self-terminates with an increase in the sinus rate, which differentiates it from complete heart block. This is important to understand when caring for ALS patients.

The presence of competing sinus and accelerated junctional rhythms can be attributed to a slowed sinus node, impaired AV conduction, or increased AV junction automaticity. Typically, etiologies of a slowed SA node include anatomic changes such as idiopathic degeneration, ischemia, medications including antiarrhythmics, and increased vagal or decreased sympathetic tone. Likewise, accelerated junctional rhythm can be caused by digoxin toxicity, inferior infarctions, myocarditis, or increased sympathetic tone at the AV junction. As both the SA and AV node are influenced by autonomic innervation, which is impaired in ALS, these rhythms could result from an imbalance in sympathetic and parasympathetic drives.

Our patient has late-stage ALS and sympathetic withdrawal may have slowed his SA node. While sinus node dysfunction with ALS has not been well delineated, a patient with ALS who

developed heart block with a junctional escape rhythm was attributed to sympathetic withdrawal and vagal predominance at the AV-node.⁴ Analysis of the relationship between ALS and various forms of heart block using EHR databases estimates heart block prevalence to be 25% higher in patients with ALS.⁴

Pseudo-Ischemic ECG Changes

As with other CNS disorders, ALS can also produce a “pseudo-infarct” pattern on ECG. In a review of 31 ECGs in ALS patients, Hindfelt et al. found only 4 patients had normal ECGs.⁶ The most common abnormality was ST-segment or T-wave changes in 16 patients, of whom 7 had no preexisting cardiovascular disease. Notably, 2 patients with ST-segment elevations were confirmed to have no evidence of myocardial ischemia. These changes were hypothesized to be driven by ALS’s effects on neuromuscular and autonomic imbalance.

Two recent case reports described ST-segment and T-wave changes in ALS. One patient who presented with chest pain with new inferior Q-waves and negative troponins, had normal coronary anatomy on cardiac catheterization.⁷ Another case reported ECG pattern of ST-elevation followed by biphasic T and inverted T-waves without detectable myocardial abnormalities.⁸ Finally, an 84-year-old woman with ALS presented with chest pain and dyspnea, with negative T-waves in precordial leads V2-6 and leads I and aVL had normal coronary arteries.⁹

Conclusions

ALS is a complex disorder with a heterogenous degree of neurologic and cardiovascular sequelae. Evidence suggests that neuromuscular and autonomic imbalance in ALS contributes to its cardiovascular effects. There is high prevalence of abnormal ECG findings and arrhythmias in ALS patients that are part of the disease process. Increased awareness of how ALS affects the cardiovascular system will allow for informed management in this progressive illness.

Figure 1: ECG-1 - Atrial flutter with 2:1 block, RBBB, possible inferior infarct pattern

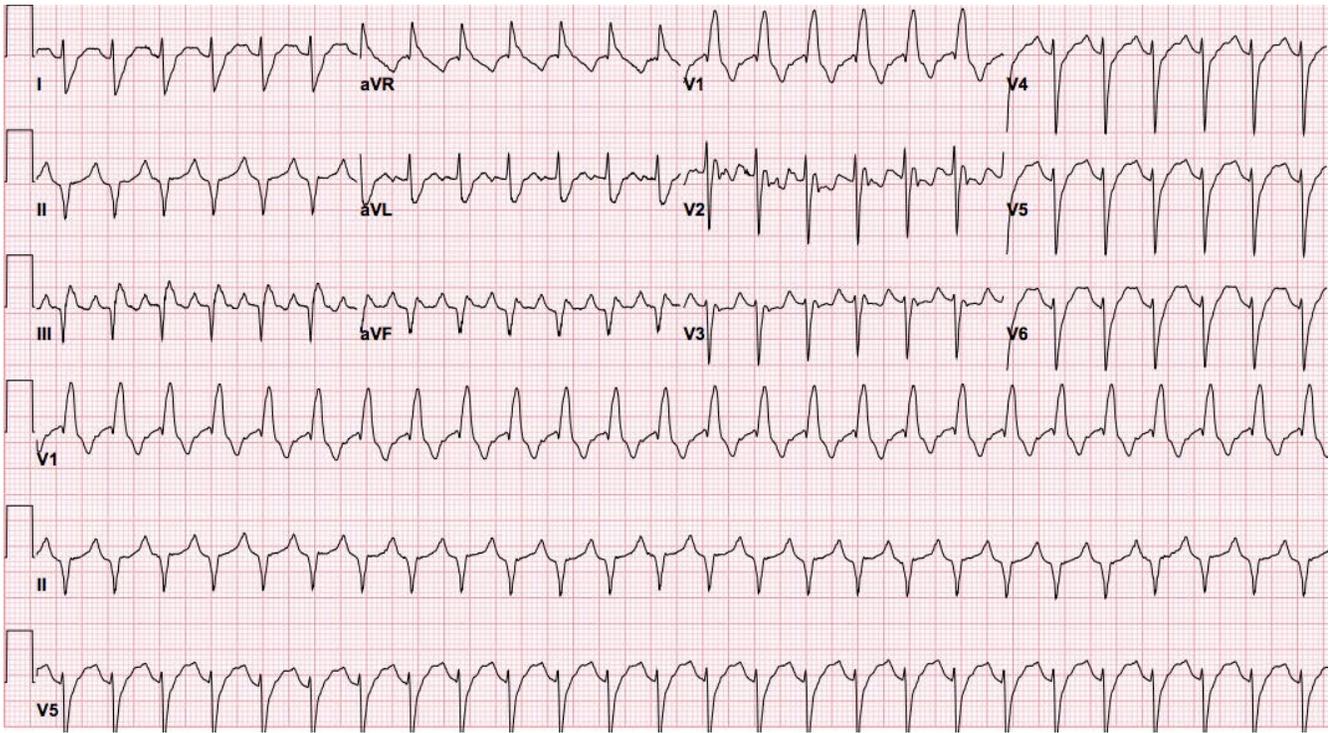


Figure 2: ECG-2 - Sinus rhythm with RBBB, LAFB with pseudo-inferior infarct pattern

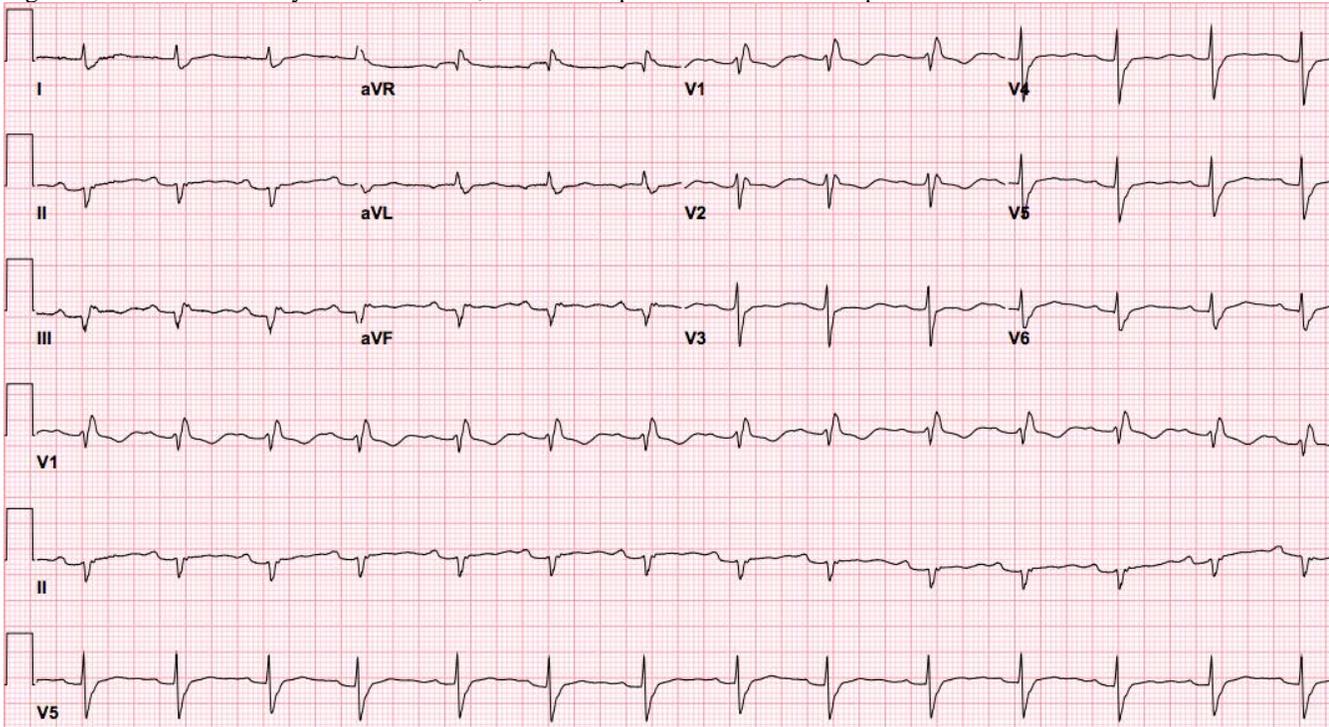


Figure 3A: Telemetry Strip A - Isorhythmic dissociation with sinus arrhythmia and pauses with atrial escape rhythm, as well as junctional escape rhythm.

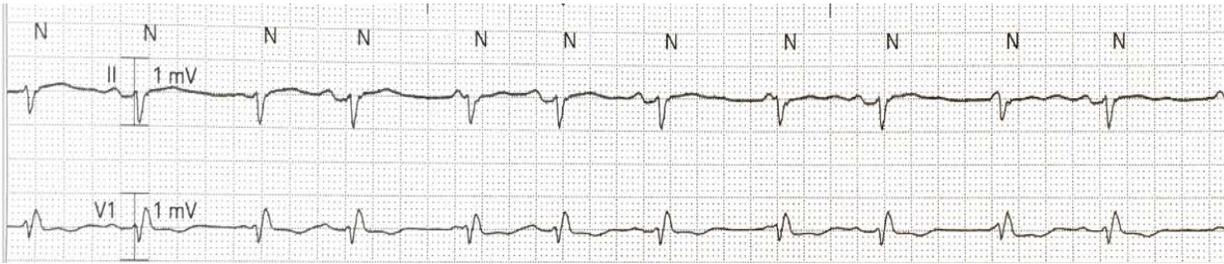


Figure 3B: Telemetry Strip B - Accelerated junctional escape rhythm

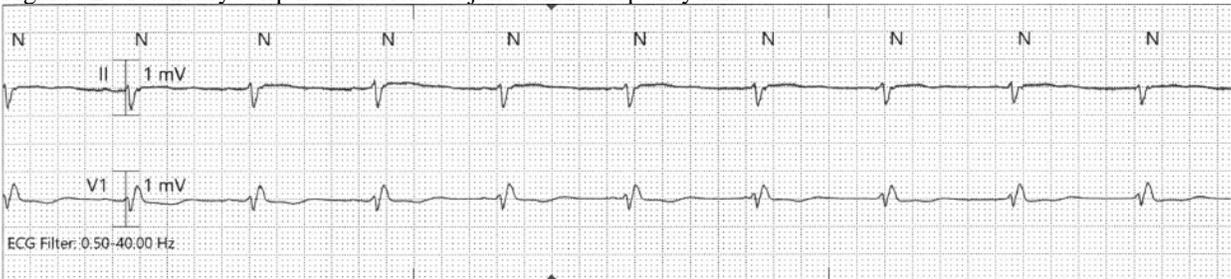
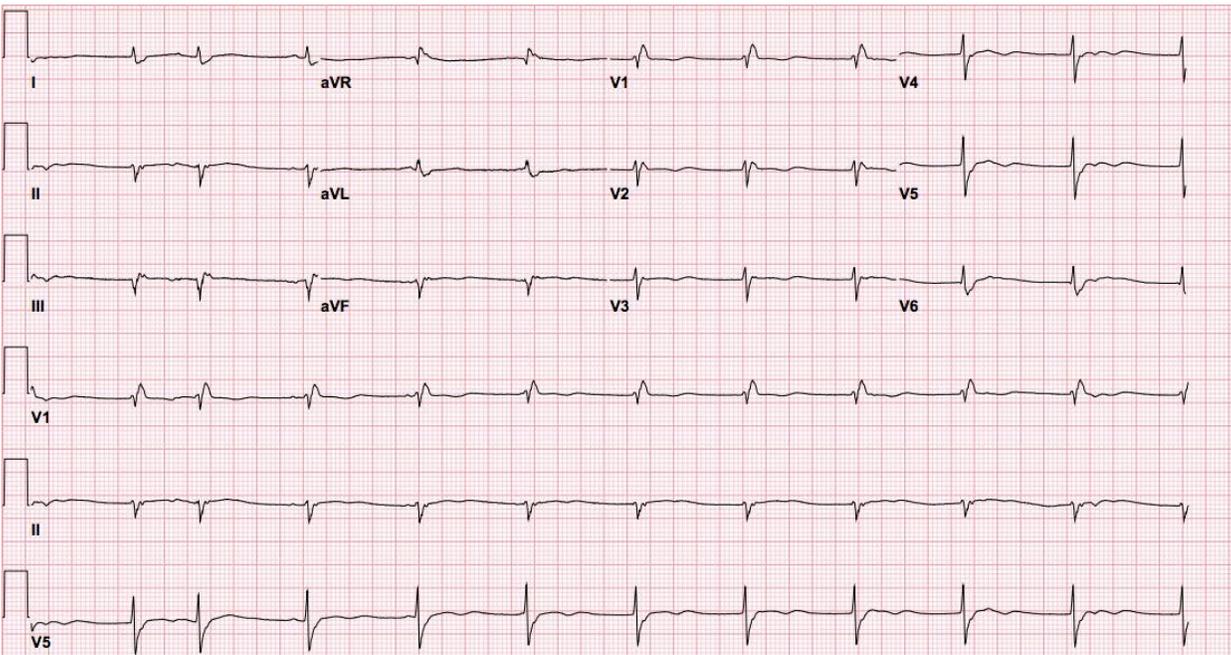


Figure 4: ECG-3 - Sinus bradycardia with isorhythmic dissociation and accelerated junctional rhythm, RBBB, LAFB with pseudo-inferior infarct pattern



Acknowledgement: This patient is a long-time educator and hopes his case can be used to educate others.

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