Thinking Outside the Cardiac Box

Anchoring in an Elderly Patient with Multiple Visits for Orthopnea

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Learning Objectives

- ☑ Recognize that neuromuscular disorders can mimic respiratory illnesses
- ☑ Include a neurological assessment in the comprehensive workup for dyspnea of unknown etiology

Case Information

HPI

77-year-old male with history of hypertension and prostate cancer presented to the emergency department four times for evaluation of progressive shortness of breath over a four-month period. Symptoms were worse with lying down and associated with fatigue.

Visit 1: Discharged from the ED with a diagnosis of dyspnea of unknown etiology.

Visit 2: Admitted to Cardiology service – negative cardiac work up. Symptoms said to be due to "deconditioning."

Visits 3 and 4: Discharged from the ED with a diagnosis of pneumonia but returned six hours later for worsening cough and dyspnea.

Physical Exam

T 36.3°C, HR 83, BP 137/74, RR 16, SpO2 94%

CV: Regular rate and rhythm. Systolic 1/6 murmur. Trace bilateral lower extremity edema.

Resp: Breath sounds normal. Accessory muscle usage present. No respiratory distress.

O2 saturation: **90% while upright**
79% while supine with severe dyspnea

Neuro: Alert and oriented to person, place and time.

Labs & Imaging

Labs:
- Respiratory viral PCR negative.
- ABG: pH 7.35, CO2 61, bicarb 32.6
- CSF normal, cytology negative
- Paraneoplastic/autoantibody, treponema, HIV negative

Cardiac Studies:
- Unremarkable ECG
- Nuclear stress test negative
- Left and right heart catheterizations normal

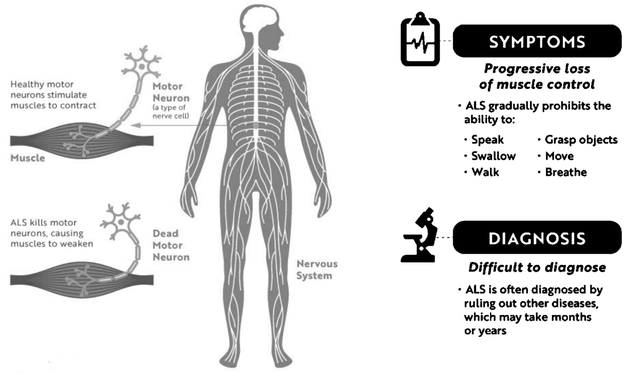
Imaging:
- CXR: Lower lobes with atelectasis vs aspiration/infection
- CT-PE: No pulmonary embolus
- TTE w/ contrast: Small right to left interatrial shunt

Hospital Course

- Admitted and worked up for pneumonia, COPD, heart failure, pulmonary shunting and dysphagia.
- Detailed physical examination: fasciculations and weakness of the upper extremities; hyperreflexia of the upper and lower extremities.
- Further discussion revealed four months of progressive weakness and several years of worsening tremors.
- Pulmonary function testing demonstrated reduced inspiratory strength.
- XR sniff test w/ fluoro: Partial right hemidiaphragmatic paresis.

Electromyography confirmed the diagnosis of amyotrophic lateral sclerosis (ALS).

Amyotrophic Lateral Sclerosis (ALS)



- Amyotrophic Lateral Sclerosis (ALS): fatal motor neuron disease which causes upper and lower motor neuron loss leading to loss of motor control and subsequent loss of activities such as eating, moving and breathing[4].
- Sporadic ALS accounts for 90-95%; Familial ALS 5-10% of all ALS cases[4].
- Most common in white males[5].

Diagnosis & Treatment

- Diagnosis is confirmed by:
  - Neurological exam[3].
  - Diagnostic tests: EMG, nerve conduction study, MRI, lab tests, LP, biopsy[3].
- Treatment is limited to life-prolonging treatment as no curative therapies exist[3].

Discussion

- Dyspnea and fatigue are frequent complaints in outpatient settings.
- Though often due to cardiac dysfunction, the differential diagnosis for true orthopnea is limited and must include neuromuscular disease.
- This case illustrates the importance of maintaining a broad differential and considering a neurologic etiology for dyspnea[1].
- This case demonstrates the wide variation in age of presentation of ALS and the need for a wide differential diagnosis for elderly patients[2].
- Although ALS has no cure to date[3], misdiagnosis can lead to delayed treatment, over-testing, reduced quality of life, and emotional distress for patients and their families.

References

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6. Images adapted from: ALAS http://www.alsa.org/news/public-awareness/als-awareness-month/2016/what-is-als.html

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