An Unusual Presentation of Rapidly Progressive Dyspnea: A Case Study

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NC APTA Fall Conference 2021

Objectives

1. Recognize early symptoms and common targets of hypereosinophilic syndrome
2. Discuss common medical management and complications related to hypereosinophilic syndrome
3. Identify areas of intervention that physical therapists could apply across various outpatient settings

Meeting the Patient: Evaluation Day

- 48-year-old Female
- Referred to cardiopulmonary physical therapy for “severe asthma vs autoimmune disease”
- Recent hospitalization for her complex presentation and severe dyspnea at rest

Subjective

- February
  - Baseline - her usual state of health
- March
  - Developed oral ulcers and sores, possibly related to previous diagnosis of lichen planus though she reports these seemed different. Also started experiencing dysphagia and odynophagia causing her to decrease her PO intake.
- April
  - Developed severe lower back pain radiating to left leg associated with numbness and foot drop. Given a course of steroids with good response. Had an EGD with biopsy at this point which showed esophageal stricture but no evidence of eosinophilic esophagitis on biopsy.
- August
  - Developed sinusitis with congestion and rhinorrhea. Prescribed a course of Augmentin and Allegra, which did not help. Started noticing skin changes with her fingers and toes becoming red and tender. Also noticed a rash on her right calf. Biopsies of this were taken which showed perivascular dermatitis with eosinophilia; however, the details of the biopsy are not know beyond this. Her absolute eosinophil count at this time was found to be 1100.
- October
  - Developed acute onset shortness of breath with walking. She was no sit down to catch her breath. She went to the ED where her workup was unremarkable and she was discharged on 10L and admitted with concerns for adult onset asthma.
- November
  - Started on Prednisone and Celebrex by her PCP for persistent wheezing. Seen by pulmonology where a 6 minute walk test was significant for SpO2 drop from 97-91. PFTs showed greatly reduced FEV1.
- December
  - Went back to the ED for shortness of breath. Was found to have SpO2 of 80%. Workup again unremarkable. Discharged home on 2L O2 and referred to pulmonary rehab.

Medical History

- Chronic sinusitis
- Low back pain (history of L4-L5 discotomy)
- History of right total hip arthroplasty
- Oral ulcers
- Dysphagia
- Recurrent rash
- Hypoxic respiratory failure
- Severe asthma vs autoimmune lung disease
Medications
Albuterol Inhaler
Aspirin
Atorvastatin (Lipitor)
Celebrex (Celecoxib)
Vitamin D3
Fluticasone propionate-salmeterol (Advair Diskus)
Furosemide (Lasix)
Albuterol nebulizer solution
Omeprazole (Prilosec)
Prednisone (Deltasone)
Sulfamethoxazole-trimethoprim (Bactrim DS)

Psychosocial Assessment
- Prior level of function: Independent
- Current level of function: Modified independent with some ADLs and iADLs
- Home environment: 1-story home with her husband
- Home DME: None
- Vocational history: currently on medical leave as a school teacher
- Exercise/Rehab history: walks around the house for exercise since hospital discharge

Assessment Continued
- General cognition, attention span, memory, orientation all intact
- Safety judgement and insight to deficits normal
- No barriers to learning
- Restricted
- No fall history
- Risk factor could be her generalized weakness

Cardiopulmonary Subjective
- Regular diet
  - No restrictions, not diabetic
- No baseline cough; does not need regular airway clearance
- No current supplemental oxygen use
- Has never smoked cigarettes
- Greatest difficulties: walking and showering
- Patient goal: "Be able to walk from parking lot to classroom without needing to rest"

Objective Examination
- Vital signs WNL
- Breathing ambient air, placed on continuous pulse oximetry throughout session
- Denies any pain
- General observation
  - Pleasant female, NAD, presents with her husband
  - Eye contact, oriented,Lisa/leads
  - Pulse WNL
- Range of Motion:
  - KNEE, LLE, RLE, and LLI: Normal ROM and not painful

Objective
- Strength:

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**Objective**

- **Sensation:**
  - LE WNL
  - LE chronic L foot numbness due to previous back surgery
- **Flexibility:** WNL
- **Posture:** no obvious structural impairment
- **Gait:** decreased cadence
- **Transfers:** Independent sit to stand, sit to supine/supine to sit
- **Balance:** able to maintain tandem stance for 10 seconds with moderate postural sway
- **Breathing pattern:** limited diaphragmatic movement; good demonstration of pursed lip breathing
- **Auscultation:** Heart - normal S1, S2; Lung - BUL wheezing

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**Objective**

- **Short Physical Performance Battery (SPPB)**
  - Balance: 4
  - 4m gait: 3 (5.65 sec)
  - 5 STS: 2 (15.57 sec)
  - Total: 9/12 points

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**Objective**

- **6-Minute Walk Test (6MWT)**
  - Rest:
    - SpO2 99%
    - HR 106, BP 110/70
  - Exercise:
    - SpO2 92%
    - HR 126, BP 140/80
    - 2 rest breaks (1 sit, 1 stand)
    - No AD
    - 728 feet
    - 39.35% Predicted

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**Objective**

- **UCSD Shortness of breath (SOBQ):** 92
- **Shortness of Breath**
  - 120 total points
  - Higher the score = higher self-reported SOB
- **Ferrans & Powers: pulmonary version (QOL):** 19.62
  - Quality of life
  - 30 total points
  - Higher the score = higher self-reported QOL
- **Center of Epidemiology Studies Depression scale (CESD):** 17
  - Depression screen
  - 60 total points
  - Score > or = 16 is at risk for depression like symptoms

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**Assessment**

- **Primary impairments:**
  - Decreased strength, limited muscle endurance, decreased functional level, dyspnea on exertion, and lack of home exercise program
  - **6MWT**
    - 728 ft (39.35% predicted); needed two rest breaks, lowest SpO2 reading was 92%
  - **SPPB**
    - 9/12, mild limitations to general mobility and increased risk for falls based on 5STS
- **Rehab enhancers:**
  - Age, good family support/resources, good insight into disability, good problem solving skills, insurance coverage, intact cognition, motivation, and previous level of function/active lifestyle
- **Rehab barriers:**
  - Low activity tolerance and medical complexity/co-morbidities

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**Goals**

- **Personal goal:** To be able to walk into her school building from the parking lot without resting
- **Rehab goals:**
  1. Walk for a total of 20 minutes without a rest break.
  2. Perform independent diaphragmatic and pursed lip breathing without verbal or tactile assistance.
  3. Independent with home exercise program along with appropriate precautions.
  4. SPPB will increase by 2 points.
  5. Patient will meet with program counselor to address quality of life and coping mechanisms.

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**Plan**

- **Frequency and Duration**
  - Decided to see her 2_ days per week for 8_ weeks
- **Aerobic exercise duration**
  - 20-60 min/session
- **Intensity**
  - RPE or OBE 3-5/10 maintaining oxygen saturation of 88-95% or greater
- **Planned interventions**
  - Aerobic/insurance exercises, breathing retraining, functional training, home exercise program, neuromuscular re-education, patient education, postural exercise, strengthening, and transfer training.
Treatment Sessions

Session 1
- Vital Signs:
  - No pain, no new reports
  - SpO2: 94%
  - HR: 112 bpm
  - BP: 108/60 mmHg
- Track walking: 14 minutes; 5 laps (1320'); no AD; 3 rest breaks; 90%, 134 HR, RPE 7/3
- Weight training: upper and lower body
- Stationary biking: 10 minutes; Level 1; 1.11 miles; 90%, 131 HR, RPE 5/3

Session 2
- Vital Signs:
  - No pain, reports tightness in her throat
  - SpO2: 94%
  - HR: 97 bpm
  - BP: 104/67 mmHg
- Track walking: 14.5 minutes; 8 laps (2112'); no AD; 1 rest; 95%, 134 HR, RPE 6.5/3
- Weight training: upper and lower body
- Stationary biking: 12 minutes; L1; 1.35 miles; 95%, 132 HR, RPE 6/3

Week 2
- Vital signs stable, especially oxygen levels on room air
- Continuous monitoring of SpO2 and HR throughout sessions
- Reported tightness in her chest on arrival both days
- Track walking: decided to do lap goals instead of setting time, also began using rollator for energy conservation
  - 7 laps (1540') in 24 minutes; 2 rest breaks; RPE 6/3
  - 9 laps (2376') in 20 minutes; 1 rest break; RPE 6/3
- Weight training: upper and lower body
- Stationary biking:
  - 10 minutes on level 1; RPE 6/3 both days

Week 3
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout sessions
- Reported increased chest tightness with increased dyspnea
- Breathing treatment
- Contacted pulmonologist for follow-up
- Track walking: deferred
- Weight training: upper and lower body modified
- Stationary biking:
  - Third upright: 10 minutes on level 1; RPE 6/3

Week 4
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout sessions
- Followed up with MD; changed inhalers, prednisone taper
- Tired, has thrush
- Track walking:
  - 8 laps (2112') in 28 minutes; RW; 1 rest break; RPE 6/3
- Weight training: upper and lower body
- Stationary biking:
  - Lied upright: 10 minutes on level 1; RPE 6/3
Week 5
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout sessions
- Reported extension of medical leave
- Track walking with rollator:
  - 9 laps (2376') in 20 minutes; NO RESTS; RPE 7/3
  - 8 laps (2112') in 20 minutes; 1 rest; RPE 7/3
- Weight training: upper and lower body
- Stationary biking:
  - 15 minutes on level 1; RPE 6/3 each day

Week 6
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout session
- Reported still having thrush
- Purchased a stationary bike for home use
- Track walking: Reassessment day "c"
  - 6MWT 55’
  - SPPB: 12/12
- Weight training: upper and lower body
- Stationary biking:
  - 10 minutes on level 1; RPE 7/3

Reassessment Comparison
Initial Evaluation 6MWT
- Rest:
  - 99% SpO2
  - 106 HR
  - 110/70 BP
- Exercise:
  - 92% SpO2
  - 126 HR
  - 140/80 BP
  - 2 rest breaks (1 sit, 1 stand)
  - No AD
  - 728 feet
  - 39.35% Predicted

10th visit 6MWT
- Rest:
  - 93% SpO2
  - 117 HR
  - 110/70 BP
- Exercise:
  - 95% SpO2
  - 125 HR
  - 120/82 BP
  - 2 rest breaks (seated)
  - No AD
  - 656 feet
  - 35.99% Predicted

Week 7
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout sessions
- Reported ongoing mouth sores and tightness in her throat
- This week she met her personal goal of walking into her school building without needing to stop!
- Track walking with rollator:
  - 8.5 laps (2244') in 20 minutes; 2 rests; RPE 7/3
  - 10 laps (2640') in 20 minutes; NO RESTS; RPE 6.5/3
- Weight training: upper and lower body
- Stationary biking:
  - 15 minutes on level 1; RPE 6/3

Week 8
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout sessions
- Bought a scooter for community mobility as needed
- Track walking with rollator:
  - 9 laps (2376’) in 20 minutes; 1 rest; RPE 7/3
  - 8 laps (2112’) in 20 minutes; 1 rest; RPE 6/3
- Weight training: upper and lower body
- Stationary biking:
  - completed at home

Week 9: Discharge
- Vital signs stable
- Continuous monitoring of SpO2 and HR throughout session
- GIVE A DIAGNOSIS!
- Track walking with rollator:
  - 8 laps (2112’) in 20 minutes; NO RESTS; RPE 7/3
  - 6MWT 55’
  - SPPB: 12/12
- Weight training: upper and lower body
- Stationary biking:
  - At home
Rehab Outcomes: 6MWT

**Initial Evaluation**
- Rest:
  - 93% SpO2
  - 117 HR
  - 110/70 BP
- Exercise:
  - 91% SpO2
  - 120 HR
  - 120/80 BP
  - 2 rest breaks (1 sit, 1 stand)
  - No AD
  - 728 feet
  - 35.99% Predicted

**Final visit**
- Rest:
  - 92% SpO2
  - 106 HR
  - 110/70 BP
- Exercise:
  - 95% SpO2
  - 127 HR
  - 122/80 BP
  - No rests
  - Rollator
  - 852 feet
  - 46.92% Predicted

**10th visit**
- Rest:
  - 99% SpO2
  - 106 HR
  - 110/70 BP
- Exercise:
  - 92% SpO2
  - 126 HR
  - 140/80 BP
  - 2 rest breaks (1 sit, 1 stand)
  - No AD
  - 728 feet
  - 39.35% Predicted

Rehab Outcomes: SPPB

**Initial Evaluation**
- Balance: 4
- 4m gait: 3 (5.65 sec)
- 5 STS: 2 (15.57 sec)
- Total: 9/12 points

**Final visit**
- Balance: 4
- 4m gait: 4 (4.93 sec)
- 5 STS: 4 (9.34 sec)
- Total: 12/12 points

**10th visit**
- Balance: 4
- 4m gait: 4 (4.93 sec)
- 5 STS: 4 (9.39 sec)
- Total: 11/12 points

Rehab Outcomes: Questionnaires

- SOBQ: 92/120
- QOL: 19.62/30
- CESD: 17.68

Finding a Diagnosis
Hypereosinophilic Syndrome

Breaking it down...

Hypereosinophilic Syndrome (HES)
- A rare group of conditions that are associated with eosinophilia with evidence of organ involvement.
- Any organ system can be involved.
- Characterized by:
  - Absolute eosinophil count >1500/uL.
  - Elevated levels lasting longer than 6 months.
  - Lack of evidence for parasitic, allergic, or other known causes of eosinophilia.
  - Signs and symptoms of eosinophil-mediated organ dysfunction.
- Eosinophil: type of white blood cell that is a granulocyte. It releases granules of enzymes to fight foreign substances and infections.
- Two important functions related to the immune system:
  - Kill invading germs such as parasites, bacteria, or viruses.
  - Promote inflammation to help the body isolate and control a disease or allergic reaction.
Breaking it down...

- High eosinophil counts could indicate your body trying to fight a bacteria, virus, or parasite; indicative of an infection
- Examples:
  - Churg Strauss syndrome: also eosinophilic granulomatisis with polyangiitis; characterized by asthma, increased eosinophils, and vasculitis
  - Allergic disorder: such as asthma, eczema, hay fever, or allergies to substances or medications
  - Autoimmune disorders
  - Blood disorders: for example leukemia

Eosinophilia

- Usually diagnosed when an absolute eosinophil count is >500/uL
- Two types
  - Tissue eosinophilia - high levels of eosinophils at a specific site of infection or inflammation (bloodstream levels are normal)
  - Blood eosinophilia - high levels through the entire bloodstream
- All of this can be detected by a blood test; tissue eosinophilia can be found in mucus, fluid, or tissue samples
- An absolute eosinophil count >1500/uL = hyper eosinophilia

Diagnosis & Treatment

- Diagnosed by lab tests and imaging
  - Blood tests, allergy tests, stool tests, or genetic tests
  - X-ray, CT scan, Echocardiogram, or MRI
- Treatment is aimed at reducing eosinophil count to prevent tissue damage
  - Specific to patient symptomology, severity of condition, and cause
  - Medication
    - Systemic corticosteroids; Imatinib or Nucala (FDA approved)
  - If all else fails, bone marrow or stem cell transplant

Prognosis

- Depends on the organ system(s) involved, disease severity, and response to therapy
- Can vary from one person to another
- No cure
- If untreated it can become life threatening

Our Patient Recap

- Symptoms
  - Ulcers
  - Dysphagia and odynophagia
  - Sinusitis and congestion
  - Low back pain with LLE numbness and foot drop
  - Skin changes/rashes
  - Dyspnea
  - Wheezing
  - Hypoxemia

Patient Medications

- Albuterol inhaler
- Aspirin
- Atorvastatin (Lipitor)
- Citalopram (Celexa)
- Vitamin C2
- Prednisone (Deltasone)
- Sulfamethoxazole-trimethoprim (Bactrim SS)

37 38 39 40

41 42
Treatment after diagnosis

- Her doctors were going to run more labs and tests
- Start Nucala (mepolizumab)
  - Injection medication for people with severe eosinophilic asthma
  - Specifically targets eosinophils
  - Helps prevent severe asthma attacks
  - Reduces need for oral steroids
  - Improves breathing
  - Given every 4 weeks

Patient Update

- Patient was walking at home and completing video exercises with bands
- Took about another month after diagnosis to get approval for new medication
  - Nucala
- Unfortunately, 2 months after completion of PT she was re-hospitalized
  - Found to have multifocal pneumonia
  - Treated for a week inpatient before discharge home

Patient Update

- Contacted patient to see if she would like to reenroll in pulmonary rehab at our center
- Referred to cardiopulmonary rehab near her hometown
  - Began about 3 weeks after discharge from hospital
  - 3 days per week for 12 weeks
  - Worked on aerobic/endurance training and strengthening

Patient Update

- She will remain out on FMLA until November
- She has the option to work from home some days and with modifications in person
- Her husband has continued to be supportive of her needs and healthcare
  - Has her personal scooter and rollator as needed
  - Able to live modified independent

Patient Update

- Work
  - Large, is being treated with the right medication now, has improved her overall stamina, known how to better manage her disease
  - Negatively, is still continuing to have dyspnea on exertion
- Overall
  - Positive: is being treated with the right medication now, has improved her overall stamina, knows how to better manage her disease
  - Negative: is still continuing to have dyspnea on exertion

Conclusions

- Patients can present with dyspnea for many different reasons
  - This patient wasn’t responding to conventional treatments
  - Condition worsened which led to further testing
  - Ultimately got a diagnosis almost a year later
  - If we had seen her sooner, would we have changed the outcome?
    - Most likely not with the progression of the disease itself
    - Could have provided communication between providers of ongoing symptoms
  - Exercise is beneficial for improving dyspnea on exertion
    - At PT we can train the body to work more efficiently aside from disease processes
    - Education on breathing techniques and home exercise can benefit patients in managing their symptoms

Questions

Thank you for your time!
Contact Information

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References


