Vasculitis and your Breathing

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OUTLINE

• Respiratory symptoms in patients with vasculitis.
  ➢ What are we talking about here?

• What can cause shortness of breath in patients with.
  ➢ A crash-course in (patho)physiology

• What your physicians can do to find out why you feel short of breath.
  ➢ Components of a systematic diagnostic work-up
Respiratory Symptoms in Vasculitis

• Shortness of breath = dyspnea (not to be confused with fatigue, malaise, weakness)
• Cough (dry, hacking, or productive of phlegm)
• Coughing up blood = hemoptysis
• “Wheezing”
  ➢ True wheezing in asthmatics, EGPA
  ➢ Stridor from subglottic stenosis, GPA
Potential Causes of Respiratory Symptoms in Vasculitis Patients

- **Active disease** affecting airways, lungs, heart, kidneys, vascular system
- **Damage** from previously active disease
- **Treatment complications** (infections, pulmonary drug toxicity)
- **Other problems** that may or may not be related to GPA and/or its treatment (obstructive sleep apnea)
Dyspnea = Shortness of Breath
oxygen supply to tissues falls behind the demand

Problems in many different organs can be the cause:

- Blood (anemia)
- Heart
- Airways
- Lung tissue
- Pulmonary vessels (thromboembolism, pulmonary HTN)
- Respiratory muscles
- Kidneys (anemia, fluid overload)
- Deconditioning after chronic illness
How System Gets Enough Oxygen

The Components of the Machinery

Oxygen Transport

$O_2$ in solution in plasma

$0.003 \text{ ml } O_2/100 \text{ ml plasma/mm Hg } P_0_2$

$O_2$ combined with $Hb$

$1.34 \text{ ml } O_2/\text{g Hb}$

Alveoli of lung

Bloodstream

Plasma

Body tissues

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Diagnostic Procedures to Identify the Cause(s) of Respiratory Symptoms in Vasculitis Patients

- CBC (Hemoglobin)
- Imaging (CXR, CT)
- Pulmonary function testing
- Oximetry, arterial blood gases
- Bronchoscopy
- ECHO-cardiography
- Cardiac stress test
- Cardio-pulmonary exercise study
- Polysomnography

Increasing Invasiveness And/or Complexity
Diffuse Alveolar Hemorrhage
What it looks like on X-ray and CAT scan
Pulmonary Function Testing

Forced Expiratory Vital Capacity Maneuver

Patient inspires maximally to total lung capacity, then exhales into spirometer as forcefully, as rapidly, and as completely as possible.
Pulmonary Function Testing

- **Lung volumes** - lung tissue loss, respiratory muscle weakness
- **Air flow** - airway obstruction, level of airway obstruction
- **Maximal inspiratory and expiratory pressures** - neuro-muscular problems
- **Oximetry with or without exertion** - is oxygenation adequate, or how much oxygen supplementation is needed?
- **Diffusion capacity** - lung tissue, lung vessels
53 y.o. Male  
WT: 130.7 KG, BMI: 35.4**  
Ht: 192.2 cm  

Previous test:  Desk:  

<table>
<thead>
<tr>
<th>LUNG VOLUMES</th>
<th>NORMAL</th>
<th>RANGE</th>
<th>CONTROL FOUND %PRED.</th>
<th>POST-DILATOR FOUND %CHANGE</th>
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<tr>
<td>TLC (Pleth)</td>
<td>7.92</td>
<td>&gt;6.55</td>
<td>8.84</td>
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<tr>
<td>VC</td>
<td>6.00</td>
<td>&gt;5.16</td>
<td>4.05*</td>
<td>68%</td>
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<tr>
<td>RV</td>
<td>1.91</td>
<td>&lt;2.49</td>
<td>4.79*</td>
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<tr>
<td>RV/TLC</td>
<td>24.2</td>
<td>&lt;31.7</td>
<td>54.1*</td>
<td>224%</td>
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<tr>
<td>FRC</td>
<td></td>
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| SPIROMETRY    |        |       |                      |
|---------------|--------|-------|----------------------|---------------------------|
| FVC           | 6.00   | >5.16 | 3.96*                | 66%                       |
| FEV1          | 4.63   | >3.95 | 1.45*                | 31%                       |
| FEV1/FVC      | 77.1   | >67.9 | 36.6*                | 45.7*                     |
| FEF25-75      | 3.9    | >2.1  | 3.5*                 | 13%                       |
| FEFmax        | 10.2   | >6.8  | 6.0*                 | 6.4*                      |
| MVV           | 168    | >135  | 72*                  | 43%                       |

| DIFFUSING CAPACITY |        |       |                      |
|---------------------|--------|-------|----------------------|---------------------------|
| DLCO(SB)            | 32.4   | >24.4 |                      |                           |
| DLCO(adjusted for Hgb = 15.4 gm/dL) | 7.63 | >6.55 |                           |
| VA                  |        |       |                      |                           |

| OXIMETRY          |      |       |                      |
|-------------------|------|-------|----------------------|---------------------------|
| O2 Sat            | 95   | > 93  | 95                   |                           |
| Pulse             |      |       | 93                   |                           |

*Outside normal range.  ** weight exceeds 98th percentile.  
**Bronchodilator was Albuterol

Asthma in EGPA
Bronchoscopy

- Inspect
- Sample
- Manipulate
GPA

Subglottic Trachea

ACTIVE DISEASE

Courtesy: UBS Prakash
GPA
Subglottic Stenosis

Vocal Cords
Expiration

Vocal Cords
Inspiration

Subglottis
Inspiration

DAMAGE

Courtesy: UBS Prakash

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Fixed Airway Obstruction
Subglottic Stenosis in WG
Endobronchial Involvement of WG
Intrathoracic Large Airway Obstruction in WG

![Graph showing maximal expiratory and inspiratory flows with predicted, control, and post-dilator curves.]
Diffuse Alveolar Hemorrhage
CXR on Admission
Treatment Complications

- **Infections**
  - Bacterial
  - Mycobacterial
  - Fungal
  - Viral

- **Drug Toxicity**
  - Methotrexate
  - Cyclophosphamide

Can mimic relapse of the underlying vasculitis
Methotrexate-associated Lymphoproliferative Disorder
3 months after discontinuation of MTX
Pulmonary Hypertension
High Blood Pressure in Pulmonary Circulation

- Acute onset or chronic
- Normal CXR, low diffusing capacity on PFT
- ECHO-cardiogram - estimate of pressure
- CT-angiogram (if pulmonary embolism suspected)
- Ventilation - Perfusion scan
- Right heart catheterization - “true numbers”, treatment decisions
Pulmonary Hypertension
High Blood Pressure in Pulmonary Circulation

- Thromboembolism (common in ANCA-associated vasculitis)
- Vascular narrowing & remodeling (common in Takayasu’s)
- Secondary = due to hypoxemia, obstructive sleep apnea a. o.
Pulmonary Embolism

Infarct in L. lower lobe. Pleural exudate over lesion.
Obstructive Sleep Apnea
Signs & Symptoms

- Morning headaches
- Daytime sleepiness (driving, watching TV, conversation etc.)
- Loss of libido
- Bad dreams
- Snoring
- Apnea spells ("choking on tongue")
Obstructive Sleep Apnea
Diagnosis

• Overnight oximetry
• Polysomnography (complete sleep study)

CPAP can turn your life around!
Summary

• There are many reasons for respiratory symptoms in vasculitis patients.
• The causes of these need to be determined in an organized fashion.
• Active disease needs to be differentiated from “damage” and from treatment complications (infections & others).
• Reversible or treatable causes need to be remedied.