Cryptogenic organizing pneumonia presenting at hypoxemic respiratory failure

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Chief Complaint: Cough and Altered mental status

History of Present Illness:
The patient is a 30 y/o male with history of developmental delay secondary to prematurity who presented to his primary care physician for five days of non-productive cough. He was thought to have atypical pneumonia vs bronchitis and he was prescribed azithromycin. After three days of therapy he was not improving and developed increased agitation. His mother brought him to the ER for further evaluation. His mother is his primary caregiver and he attends an adult day care program three days per week. Prior to starting the azithromycin he required no medications on a daily basis. His mother reports no fever, chills or phlegm production.

Exam:
Vitals: BP:136/77 HR:124 R:10 T:99.3 Weight: 60kg
General: irritated, in distress
HEENT: dry mucous membranes
Cardiovascular: tachycardia, no murmur
Pulmonary: tachypnea, accessory muscle use, respiratory distress, coarse breath sounds bilaterally with crackles at the bases
Gastrointestinal: soft, non-distended
Musculoskeletal: no rashes, no lower extremity edema

Labs:

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<th>Na</th>
<th>WBC</th>
<th>Blood Cx</th>
<th>Hgb</th>
<th>Urine Cx</th>
<th>Hct</th>
<th>Urine Legionella</th>
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<td>8.7</td>
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<tr>
<td>K+</td>
<td>Cl</td>
<td>Urine Legionella</td>
<td>PCO2</td>
<td>Hct</td>
<td>313</td>
<td>Urine Histoplasma</td>
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<tr>
<td>7.9</td>
<td>90</td>
<td>Neg</td>
<td>55</td>
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<tr>
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<td>Hct</td>
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<td>Urine Blastomycosis</td>
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<tr>
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<td>BAL #1</td>
<td>RBC</td>
<td>785</td>
<td>Segmented Neutrophils</td>
<td>BAL #2</td>
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Histological evidence of Mason Bodies in lung parenchyma

CT Chest 5 months post discharge

Chest radiograph & CT chest on admission

Discussion:
Organizing pneumonia is defined as an inflammatory lung disease with distinctive clinical, radiological and pathological features.

Clinical
- subacute presentation with fever, non-productive cough, malaise, weight loss, dyspnea

Radiological
- Patchy alveolar opacities with peripheral and bilateral distribution (See A marked in images)

Pathological
- Intra-alveolar buds of granulation tissue consisting of fibroblasts, myofibroblasts and loose connective tissue. (Masson bodies) See B in images

Classified as Cryptogenic or Secondary.

Organizing Pneumonia

Idiopathic

Secondary

Drugs

Infections

Connective Tissue Disease

This has previously been called bronchiolitis obliterans organizing pneumonia (BOOP) and has routinely been a histologic diagnosis. The average age of onset is 60 years old and it has an equal prevalence for men and women. Common presentation includes malaise, cough, fever and dyspnea. In a recent retrospective study in 2011 the average time until diagnosis was on average 3 months and the work-up is primarily an outpatient process. Lung biopsy is used to review the characteristic findings and look for Masson bodies (B) which is used to confirm the diagnosis. Recurrence for organizing pneumonia is about 50% within 1 year, the mortality is low (around 5%) and most cases are managed as an outpatient disease.

Unique properties in our patient:
- Rapid progression to type 1 acute respiratory failure requiring intubation
- The patient was younger than normal (30 y/o)
- Acute presentation of 5 days duration
- Concurrent acute renal failure

Conclusion of Case
- Our patient responded appropriately to corticosteroid therapy and was extubated within 24 hours. He continued to improve clinically over the following week.
- All cultures remained negative for the duration of the hospitalization
- He was discharged on a corticosteroid taper over 6 months

Key Points
- Organizing Pneumonia is a non-infectious process with bilateral patchy alveolar infiltrates on imaging
- Standard treatment includes corticosteroids with a slow wean over 6-8 months
- There are typical imaging findings and pathological findings
- Should be included in differential for patients with non-specific complaints and bibasilar infiltrates

References

2. JF, duBois RM. Cryptogenic organizing pneumonia. Chest. 139. 4. April 2011