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Be Careful What You Screen For: An Incidental Finding Of Tracheobronchial Amyloidosis

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Be careful what you screen for:
An incidental finding of tracheobronchial amyloidosis

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Lung Cancer Screening

USPSTF Guideline: Recommends “screening for lung cancer with low-dose computed tomography (LDCT) in adults aged 55 to 80 years who have a 30 pack-year smoking history and currently smoke or have quit within the past 15 years. Screening should be discontinued once a person has not smoked for 15 years or develops a health problem that substantially limits life expectancy or the ability or willingness to have curative lung surgery.”

Benefits:
- Higher cure rates.
- Less invasive resection.
- Decreased rates of smoking.

Harms:
- False positive and incidental findings.
- Further work-up: cost, stress.
- Radiation exposure
- Overdiagnosis.

Number needed to screen to prevent one lung cancer related death: 320
20% reduction in relative lung cancer mortality.
24% of LDCTs were abnormal. 96% of those, had false positive findings with 90% requiring further investigation.

Amyloid Subtypes

<table>
<thead>
<tr>
<th>SUBTYPE</th>
<th>Disease Association</th>
<th>Amyloid Protein</th>
<th>Main Systems Affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>AL Amyloidosis</td>
<td>Plasma cell dyscrasias (IMGS, multiple myeloma), Waldenstrom macroglobulinaemia</td>
<td>Monoclonal light chains, Lambda</td>
<td>Many organs potentially affected</td>
</tr>
<tr>
<td>Hereditary Amyloidosis</td>
<td>Inherited</td>
<td>Mutated transthyretin (TTR), fibrinogen α chain</td>
<td>Many organs potentially affected</td>
</tr>
<tr>
<td>AA Amyloidosis</td>
<td>Rheumatoid arthritis, inflammatory bowel disease, familial Mediterranean fever, chronic infection</td>
<td>Serum amyloid A protein</td>
<td>Mainly Renal</td>
</tr>
<tr>
<td>Age-related (&quot;senile&quot;) Amyloidosis</td>
<td>Age</td>
<td>Wild-type TTR</td>
<td>Mainly Heart</td>
</tr>
<tr>
<td>Dialysis-related Amyloidosis</td>
<td>Dialysis for any reason</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Tracheobronchial amyloidosis: Deposits in trachea and bronchi. Usually localized. AL subtype is most common. Treated symptomatically.


Diffuse Alveolar-Septal Amyloidosis: Deposits in septa and vessel walls. AL or ATTRWt subtypes are most common. Systemic. Often asymptomatic.

Take Home Points
- One lung cancer death prevented for every 320 average risk patients screened with CT (vs CXR).
- 20% of patients will have false positive findings requiring further evaluation.

Amyloidosis:
- Amyloid in the tissue requires testing for subtype & systemic disease.
- The AL amyloid subtype is associated with plasma cell dyscrasias.
- Localized tracheobronchial amyloidosis is one of three types of pulmonary amyloidosis.

Discussion
This case explores one patient’s lung cancer screening, and the >$5,000 work up that followed.

- Lung cancer screening can save lives but often with a cost. Physicians must educate patients about the risk of false positive results necessitating additional testing.
- To make a final diagnosis in this patient more testing was required: bronchoscopy, pathology, mass spectrometry, SPEP, UPEP, serum free light chains, and a fat pad biopsy.
- Her final diagnosis, localized tracheobronchial amyloidosis, is managed symptomatically with surgical treatments and stenting.

References