Diffuse Tracheal Narrowing

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Patient Presentation

- 59-year-old male with a history of chronic illness returning for appraisal of his respiratory status
- He has been followed by a pulmonologist and has received methotrexate and prednisone therapy in the past
- Review of systems and physical examination were unremarkable
- Recent labs: CBC within normal limits
  ESR 4
  BUN 16
CT Scan

High attenuation tracheal wall with thickening and luminal narrowing

Patient

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CT Scan

Normal tracheal anatomy at similar level

Patient

High attenuation tracheal wall with thickening and luminal narrowing

Comparison
CT Scan

Tracheal narrowing (11 mm) and wall thickening (4 mm)

Right main stem bronchus = 7 mm

Left main stem bronchus = 5 mm

Patient

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CT Scan

Normal diameter of intrathoracic trachea
Male: 20 +/- 2 mm
Female: 17 +/- 2 mm

Tracheal narrowing (11 mm) and wall thickening (4 mm)
CT Scan

Normal diameter of main stem bronchi:
- Right: 15 mm
- Left: 13 mm

Patient
- Right main stem bronchus: 7 mm
- Left main stem bronchus: 5 mm
Findings

- Diffuse calcification and wall thickening of trachea and main stem bronchi with associated tracheobronchial narrowing
- Tracheal narrowing most prominent below level of aortic arch
- Sparing of the subglottic region
- Unremarkable lung parenchyma
Differential Diagnosis

Let’s consider the differential diagnosis for diffuse tracheal narrowing...
Differential Diagnosis

1. Relapsing Polychondritis
2. Wegener’s Granulomatosis
3. Tracheobronchial Amyloidosis
4. Tracheobronchopathia Osteochondroplastica
5. Postinfectious stenosis
6. Sarcoidosis
7. Rhinoscleroma
8. Saber-Sheath Trachea
Diagnosis

- This patient presented with a preexisting diagnosis...
Diagnosis

- Relapsing polychondritis
Discussion

A description of relapsing polychondritis will help us understand the radiological findings in this patient...
Discussion

• Relapsing Polychondritis (RP)
  – Definition
    • Rare multisystem disorder characterized by recurrent inflammation and destruction of cartilaginous structures
  – Epidemiology
    • Men and women affected equally
    • Most commonly affects Caucasians although it has been reported in various ethnic groups
    • Rare: incidence approximately 1/250,000
    • Average age at presentation = 47 years
Discussion

- **Relapsing Polychondritis**
  - **Etiology**
    - Unknown!
  - **Pathophysiology**
    - ? Autoimmune: pathological finding of T-cells and Ag-Ab complexes + symptomatic improvement with steroid treatment
    - **Affected tissues**
      - *Cartilage* of nose, ears, joints, larynx, trachea, major bronchi
      - *Proteoglycan-rich sites* such as eyes, inner ears, blood vessels, and heart
Discussion

- Relapsing Polychondritis
  - Signs and symptoms
    - Auricular chondritis (85-91%)
    - Polyarthritis (52-85%)
    - Nasal chondritis (48-72%)
      - Saddle nose deformity
    - Respiratory tract chondritis (40-56%)
      - Poor prognostic sign
      - Tracheal tenderness
    - Cardiovascular disease (24%)
      - Valvular dysfunction
      - Aortitis/aortic aneurysm
    - Audiovestibular damage (46-50%)
    - Ocular inflammation (50-65%)

Concomitant

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Discussion

● Relapsing Polychondritis
  – Associated conditions
    ● Rheumatoid arthritis, systemic vasculitis, various connective tissue diseases
  – Diagnosis
    ● McAdam el al criteria (3 of 6 present)
      – Bilateral auricular chondritis
      – Polyarthritis
      – Nasal chondritis
      – Ocular inflammation
      – Respiratory tract chondritis
      – Audiovestibular damage
    ● Other diagnostic systems are similarly based upon inflammation of multiple cartilages and associated symptoms
Discussion

- Relapsing Polychondritis
  - CXR characteristics
    - Tracheal stenosis
    - Calcification of cartilaginous structures
    - Evidence of coexisting vasculitis (i.e. pulmonary parenchymal infiltrates)
Discussion

- Relapsing Polychondritis
  - CT characteristics
    - Increased airway wall attenuation
      (from subtle to frank calcification)
    - Increased airway wall thickness
    - Luminal narrowing of trachea and bronchi
    - Cylindric bronchiectasis
    - Air trapping
    - Airway collapse

Expiratory CT

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Discussion

- Relapsing Polychondritis
  - Other imaging modalities
    - MRI
      - Better distinction between edema, fibrosis, and inflammation
    - Bone scintigraphy
      - Useful in localizing sites for costochondral biopsy when diagnosis cannot be made clinically
Discussion

- What other conditions could have explained these findings?
- Let’s consider some other conditions that are on the differential…
Discussion

● Wegener’s Granulomatosis
  - Necrotizing granulomatous vasculitis of upper and lower respiratory tracts and kidneys
  - Clinical
    ● M > F but females most commonly affected by tracheal involvement
  - Radiologic findings
    ● Granulomatous lung nodules/masses with central necrosis and cavitation
    ● Tracheobronchial involvement (16% of cases)
      - Predominantly affects subglottic region
      - Circumferential mucosal thickening/luminal narrowing
      - Ulceration
    ● Sinus and nasal mucosal thickening
Discussion

- Wegener’s Granulomatosis

Circumferential mucosal thickening

Tracheal narrowing from subglottic region to thoracic inlet
Discussion

- **Tracheobronchial Amyloidosis**
  - Focal or diffuse deposition of amyloid in the submucosa of the trachea and proximal bronchi
  - Clinical
    - M > F
    - Symptoms usually begin in middle or late adult life
    - Usually “primary” without systemic involvement
  - Radiologic findings
    - Circumferential tracheobronchial thickening/luminal narrowing
    - Submucosal amyloid = soft tissue density
    - Focal form: airway polyps
    - Diffuse form: diffuse airway calcification/ossification
    - Pulmonary complications
      - Obstructive pneumonia, bronchiectasis
Discussion

- Tracheobronchial Amyloidosis

Mucosal thickening involving the posterior wall

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Discussion

- **Tracheobronchopathia Osteochondroplastica (TBO)**
  - Idiopathic benign disease of trachea and main bronchi characterized by multiple submucosal osteocartilaginous nodules
  - Clinical
    - 3:1 male predilection
    - Most patients asymptomatic
  - Radiologic features
    - Spares membranous posterior wall
    - Affects lower 2/3 of trachea and proximal bronchi
    - TBO vs. RP
      - Diffuse narrowing not typical in TBO
      - Characteristic clinical stigmata with RP (i.e. auricular chondritis)
Discussion

- Tracheobronchopathia Osteochondroplastica (TBO)

![Image of CT scan showing a calcified nodule with sparing of posterior wall.]

- Calcified Nodule
- Sparing of posterior wall

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Discussion

- Postinfectious stenosis
  - Infectious agents
    - Tuberculosis, histoplasmosis, fungi
  - Mechanism
    - Infectious necrosis and ulceration of mucosa leading to granulation tissue and fibrotic stenosis
  - Radiologic features
    - Multiple areas of focal stenosis
    - Calcification of stenotic regions is rare
    - Segmental or lobar atelectasis is common
Discussion

- Tuberculosis

Focal stricture of left main bronchus just distal to the carina

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Discussion

- **Saber-Sheath Trachea**
  - Fixed deformity of the intrathoracic trachea
  - Clinical
    - Exclusively affects older males with evidence of COPD
    - Tracheal deformity a reflection of chronic transmission of increased intrapleural pressure and injury from chronic cough
  - Radiologic findings
    - Tracheal coronal diameter $< 2/3$ the sagittal diameter
    - Emphysematous changes of lung parenchyma
Discussion

● Key points
  – Differentiate focal from diffuse tracheal disease
    ● Focal stenosis may be a complication of endotracheal intubation
  – TBO and RP
    ● Posterior membranous portion of the trachea usually spared
  – Tracheobronchial amyloidosis
    ● Focal or diffuse involvement but usually circumferential
  – Wegener’s Granulomatosis
    ● Most commonly affects subglottic region
  – CT findings in combination with clinical assessment are keys to accurate diagnosis
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References


The End