Lymphangitic Carcinomatosis: Some HRCT Findings

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Lymphangitic Carcinomatosis: Overview

- Also known as “lymphangiotic spread of tumors”, “lymphangitis carcinomatosa”, or “lymphangiosis carcinomatosa”

- LC refers to the infiltration of pulmonary parenchymal lymphatic channels by tumor cells

- Usually spread to lungs is hematogenous, while spread within lungs is lymphangitic

- Unilateral (primary lung cancer and breast carcinoma) involvement is less common than bilateral (Stomach, Pancreas, Prostate, Cervical, Thyroid, Colon, or Adenocarcinoma from an unknown site)

- ~80% of the metastases are adenocarcinomas

Menu of Radiological Tests

- **Chest X-ray (CXR)**
  - ~50% of lymphangitic carcinomatosis patients appear normal on CXR

- **High Resolution Computed Tomography (HRCT)**
  - Currently appears to offer the best combination of cost and sensitivity. 8-74% better than CXR for lymphangitic carcinomatosis

- **Positron emission tomography with radio-labeled [18F]-2-fluoro-deoxy-D-glucose (PET-FDG)**
  - Very expensive. Sensitivity appears to be identical to HRCT though experience with PET-FDG in diagnosing lymphangitic carcinomatosis is limited

HRCT findings in Lymphangitic Carcinomatosis

- Irregular, smooth or nodular thickening of interlobular septa
- Irregular and nodular thickening of peribronchovascular sheets
- Thickening of centrilobular structures
- Peripherally located wedge shaped densities representing edema from lymphatic destruction
- Pleural disease
- Correct first diagnosis 40% of the time

Lymphangitic Carcinomatosis: Clinical Symptoms

- The clinical symptoms often precede radiographic abnormalities and include:
  - Breathlessness
  - Dry cough
  - Hemoptysis (unusual unless there is primary lung cancer)

- Screening for lymphangitic carcinomatosis occurs in the context of prior malignancy

British Thoracic Society and Standards of Care Committee, *Thorax* 1999;54;S1-S28
Radiograph: 1-mm lung slice taken from peripheral lower lobe – showing secondary lobule

$S \rightarrow$ Lobules are marginated by thin interlobular septa
$V \rightarrow$ Pulmonary vein branches
$B \rightarrow$ Bronchioles
$A \rightarrow$ Centrilobular Pulmonary arteries
Pathology: Interlobular septal and peribronchovascular thickening in lymphangitic carcinomatosis

On cut lung surface, small white arrows point to thickened interlobular septa and large arrow points to thickened peribronchovascular interstitium

H&E specimen (10x). Black arrows point to nodules of tumor in interlobular septa & centrilobular peribronchovascular region.
Our Patient: 35 y/o woman with 2 lung nodules. Rest of history withheld for now.

NOTE: Inspecting level above and below suspected level of lesion helps distinguish between nodule and vessel.

Axial C+ CT of the chest

Images from PACS BIDMC

Courtesy Dr. Ferris
Differential for solitary nodule

- Malignancy (adenocarcinoma - 40%, squamous cell carcinoma - 20%, large cell carcinoma - 15%, bronchoalveolar carcinoma - 10%, solitary metastases)

- Benign neoplasms (hamartomas, lipomas, and fibromas)

- Vascular lesions - Arteriovenous malformation

- Infectious granulomas - Tuberculosis, atypical mycobacterial infection, histoplasmosis, coccidioidomycosis, and blastomycosis

- Other infections - Aspergilloma, ascaris, dirofilariasis, echinococcal cyst, and bacterial abscess

- Noninfectious granulomas - Rheumatoid arthritis, Wegener granulomatosis, and sarcoidosis

- Developmental lesions - Bronchogenic cyst

- Other conditions - Hematoma, bronchiolitis obliterans-organizing pneumonia, pseudotumor, pulmonary infarction, amyloidoma, rounded atelectasis, and mucoid impaction

Our Patient: At 8 mth follow up, presents with tachycardia and dyspnea

Enlarged hilum
Filling defects
Peripheral opacities (probably cellular or fluid infiltrates)

Reformatted HR CTA Axial View of the chest

Images from PACS BI DMC
Courtesy Dr. Ferris
Our Patient: Also had diffuse pulmonary disease

- Enlarged Hilum
- Thickened Septa
- Thickened centrilobar structures
- Peripheral opacities (probably fluid or cellular infiltrates)

Axial C+ CT of the chest

Images PACS BIDMC

Courtesy Dr. Ferris
Our Patient: Diffuse interstitial disease on reconstructed Sagittal C+ HRCT of the Chest

Diffuse nodular and septal infiltrates
Major fissure

Serial Slices from Reconstructed Sagittal C+ HRCT of the chest

Courtesy Dr. Ferris
Our Patient: Interstitial infiltrates on CXR and HRCT

Images from PACS BIDMC Courtesy Dr. Ferris

Scauplic
SVC Porta catheter
Mediastinal enlargement
Increased interstitial markings

Images from PACS BIDMC

Courtesy Dr. Ferris
Differential for septal and centrilobar thickening

- **Smooth interlobar septal thickening:**
  - pulmonary edema, hemorrhage, or veno-occlusive disease; Lymphangitic carcinomatosis; lymphangiomatosis; amyloidosis, pneumonia, alveolar proteinosis

- **Nodular interlobar septal thickening:**
  - Lymphangitic carcinomatosis; lymphoproliferative disease (e.g. lymphocytic interstitial pneumonia); sarcoidosis; silicosis and coal workers pneumoconiosis; amyloidosis

- **Centrilobular nodules due to perilymphatic disease:**
  - Lymphangitic carcinomatosis; sarcoidosis; silicosis, coal workers pneumoconiosis; lymphocytic interstitial pneumonia

Our Patient: Complete History

- The patient is s/p right colectomy for poorly differentiated signet cell mucinous carcinoma of the terminal ileum; b/l salpingo-oopherectomies for mets. Mets to hemidiaphragm, bladder, small bowel, and pelvic side wall also observed. She is currently undergoing chemo.

- This combined with the CT findings makes the suspicion for metastasis to the lung and for lymphangitic carcinomatosis very high.
Prognosis for Lymphangitic carcinomatosis

- Usually very poor – survival in months
- In a small 8yr study of 10 pts treated with surgical resection of primary, chemo, and/or radiation tx, initially:
  - Pulmonary Sxs regressed in 6
  - Progressed in 2
  - Unchanged in 2
  - Median survival post diagnosis was 13 mths (range: 11-30mths)

Conclusions

- Lymphangitic carcinomatosis spreads hematogenously to the lungs and then invades the lymphatic vessels.

- The accumulation of tumor cells causes thickening of the secondary lobule interstitium.

- HRCT is currently the modality of choice for screening lymphangitic carcinomatosis patients.

- The diagnosis of lymphangitic carcinomatosis requires a clinical context that includes malignancy.

- The prognosis for lymphangitic carcinomatosis is poor.
References

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