February 2014 Imaging Case of the Month

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Clinical History: A 60-year-old man presented with a history of weight loss and dysphagia for about 2 weeks duration. There was a possible history of asthma accompanied by ongoing shortness of breath first noticed nearly 2 years ago. Frontal chest radiography (Figure 1) was performed.

Figure 1. Frontal chest radiography.

Which of the following statements regarding the chest radiograph is **most accurate**?
(Choose the correct answer to move to the next panel)

1. The chest radiograph shows a mass
2. The chest radiograph shows hilar and mediastinal lymph node enlargement
3. The chest radiograph shows multifocal consolidation
4. The chest radiograph shows multifocal, somewhat basal predominant linear opacities suggesting fibrosis
5. The chest radiograph shows multiple nodules

Correct!

4. The chest radiograph shows multifocal, somewhat basal predominant linear opacities suggesting fibrosis

The predominant findings on the chest radiograph are linear and reticular opacities, with some areas showing small cystic foci. These findings are best seen in the lung bases bilaterally. No discrete lung nodules are seen and the mediastinal and hilar contours are within normal limits. No mass is evident. While there is some increased attenuation in the bases, likely the result of the summation of the linear and reticular opacities that are present, no significant consolidation is present.

Which of the following is **the least appropriate consideration** among the differential diagnostic possibilities for the appearance of the patient's chest radiograph? (Choose the correct answer to move to the next panel)

1. Asbestosis
2. Chronic aspiration
3. Desquamative interstitial pneumonia
4. Sarcoidosis
5. Usual interstitial pneumonia / idiopathic pulmonary fibrosis

Correct!
4. Sarcoidosis

Among the possibilities listed, sarcoidosis is the only disorder that is typically upper lobe predominant; the other conditions are typically either basal predominant (usual interstitial pneumonia, asbestosis, chronic aspiration) or diffuse (desquamative interstitial pneumonia) in distribution. Additionally, sarcoidosis often appears more nodular, rather than reticular or linear, although later stage sarcoidosis, when complicated by fibrosis, may have a radiographic appearance dominated by linear and reticular opacities with architectural distortion.

The patient underwent thoracic CT (Figure 2) for further characterization of the chest radiographic findings.

Figure 2. Representative static thoracic CT displayed in lung windows.

Which of the following statements regarding this CT examination is **most accurate**?
(Choose the correct answer to move to the next panel)

1. The thoracic CT shows a fibrotic process distinct from most idiopathic interstitial pneumonias
2. The thoracic CT shows features characteristic of smoking-related interstitial lung disease
3. The thoracic CT shows features strongly suggesting lymphangitic carcinomatosis
4. The thoracic CT shows findings characteristic enough of usual interstitial pneumonia / idiopathic pulmonary fibrosis that surgical biopsy may be deferred
5. The thoracic CT shows findings suggesting active infection

Correct!

1. The thoracic CT shows a fibrotic process distinct from most idiopathic interstitial pneumonias

The thoracic CT shows peripherally distributed coarse linear and reticular abnormalities associated with subpleural cystic change, the latter related to honeycombing. Some architectural distortion is present. These features are typical of fibrotic lung diseases. The presence of findings suggesting fibrosis argues strongly against lymphangitic carcinomatosis- this disorder is not characterized by fibrosis. There are no features that specifically suggest active infection, such as areas of ground-glass opacity unassociated with findings of fibrosis, consolidation, or nodules. While usual interstitial pneumonia / idiopathic pulmonary fibrosis merits consideration for the imaging findings, the distinct subpleural sparing of the extreme costophrenic angles argues against this disorder. Smoking-related disorders may show wide variety of findings at thoracic CT, including emphysema, nodules and cysts / cavities (with Langerhans cell histiocytosis), areas of ground-glass opacity which may be centrilobular, to basal reticular changes. Emphysema, nodules, cavities, and ground-glass opacity are not evident on this study, and, when reticulation occurs as a manifestation of smoking-related lung changes, that finding is typically seen in the setting of emphysema, which is lacking. The best answer among those listed is choice “1”- features of fibrosis are present, but the conspicuous extreme costophrenic angle sparing argues strongly against certain idiopathic interstitial pneumonias, such as usual interstitial pneumonia / idiopathic pulmonary fibrosis and non-specific interstitial pneumonia.

Further clinical course: Older thoracic CTs from 6 years (Figure 3), 4 years (Figure 4), and 2 years prior to presentation (Figure 5) became available for review.

Figure 3. Representative static thoracic CT displayed in lung windows from 6 years prior to presentation.

Figure 4. Representative static thoracic CT displayed in lung windows from 4 years prior to presentation.

Figure 5. Representative static thoracic CT displayed in lung windows from 2 years prior to presentation.

Which of the following statements regarding these CT examinations is **most accurate**?
(Choose the correct answer to move to the next panel)

1. The serial thoracic CT scans show non-specific findings and do not suggest a unifying diagnosis
2. The serial thoracic CT scans show features suggesting stable fibrotic lung disease
3. The serial thoracic CT scans show a fibrotic process complicated by malignancy
4. The serial thoracic CT scans show features suggesting fibrotic process complicated by a new, potentially reversible inflammatory disorder
5. The serial thoracic CT scans show findings suggesting an inflammatory disorder progressing to fibrosis

Correct!

5. The serial thoracic CT scans show findings suggesting an inflammatory disorder progressing to fibrosis

The thoracic CT 6 years prior to presentation (Figure 3) shows inhomogeneous lung opacity, likely due to small airway obstruction, given the lobular configuration. The subsequent CTs show these same findings, but with the additional development of areas of ground-glass opacity and fine linear and reticular opacities (Figures 4 and 5), eventually progressing to features consistent with fibrosis (Figure 2); this sequence suggests an inflammatory disorder progressing to fibrosis. The serial CT scans clearly show a progressive abnormality, not a stable one. Fibrotic lung disease may be complicated by malignancy, but when this occurs, thoracic CT will commonly show multifocal or diffuse fibrotic-appearing abnormalities with a new or growing lung nodule or mass, possibly new or increasing lymph node enlargement, and such features are not present in this case. When fibrotic lung disease is complicated with a new inflammatory disorder, as may occur in patient with usual interstitial pneumonia / idiopathic pulmonary fibrosis with accelerated interstitial pneumonia, thoracic CT often shows areas of new ground-glass opacity and/or consolidation superimposed on features of fibrotic lung disease, such as reticulation, linear opacity, traction bronchiectasis, architectural distortion, and honeycombing; this sequence is more or less the reverse of the findings illustrated in the images presented.

A more focused history was taken. Which of the following would be **the most likely relevant historical point** discovered? (Choose the correct answer to move to the next panel)

1. The patient has an esophageal disorder
2. The patient has had a stem cell transplant
3. The patient is a long-time heavy smoker
4. The patient owns two parakeets
5. The patient works as a sandblaster

Correct!

4. The patient owns two parakeets

The historical point that the patient works as a sandblaster would suggest the possibility of silicoproteinosis, which, at thoracic CT appears identical to pulmonary alveolar proteinosis (PAP). PAP often appears as “crazy paving” at CT- multifocal ground-glass opacity associated with interlobular septal thickening and intralobular lines, frequently associated with a sharp demarcation between normal and abnormal lung- but that pattern is not present on the CT scans presented. The historical point of long term, heavy smoking suggests the possibility of smoking-related lung disorders, which could have multiple manifestations at thoracic CT, including upper lobe nodules / cavities (Langerhans cell histiocytosis), ground-glass opacity centrilobular nodules and/or areas of multifocal or diffuse ground-glass opacity (respiratory bronchiolitis-interstitial lung disease and desquamative interstitial pneumonia), and basal, posterior, subpleural ground-glass opacity and reticulation (smoking-related fibrosis), but often these findings are associated with emphysema, which is absent on all the CT scans presented. The historical point of stem cell transplant would suggest the possibility of constrictive bronchiolitis as a manifestation of graft-versus-host disease. The earliest CT (Figure 3), perhaps the CT obtained 4 years prior to presentation (Figure 4) as well, could be consistent with that diagnosis, given that the presence of lobular low attenuation on these scans suggests the presence of small airway obstruction. However, areas of ground-glass opacity were developing even at the time of the scan obtained 4 years prior to presentation (Figure 4), and later CTs, including the presentation CT, clearly show progressive fibrotic lung disease, which is not typical for graft-versus host disease. The historical point that that patient may have an esophageal disorder should raise the possibility of aspiration. The progressive fibrotic abnormalities in the bases is potentially consistent with that consideration, but the basal sparing present on the presentation CT (Figure 2) and the additional features suggesting small airway obstruction suggest an alternative consideration. The historical point that the patient owns two parakeets suggests the possibility of hypersensitivity pneumonitis, and the imaging sequence presented is very suggestive of that disorder.

Which of the following **should be performed** to establish the diagnosis for this patient? (Choose the correct answer to move to the next panel)

1. Bronchoscopy with bronchoalveolar lavage
2. Surgical lung biopsy
3. Transthoracic fine needle aspiration biopsy
4. 1 or 2
5. 2 and 3

Correct!

1. Bronchoscopy with bronchoalveolar lavage

2. Surgical lung biopsy

4. 1 or 2

Transthoracic fine needle aspiration biopsy is generally reserved for indeterminate focal lung disorders, such as lung nodules or masses, and not diffuse lung diseases. Usually surgical lung biopsy is required for the diagnosis of indeterminate fibrotic lung disorders, and bronchoscopy with bronchoalveolar lavage is employed principally used to exclude competing diagnoses such as infection. However, in this circumstance, with a clinical history clearly identifying an exposure and serial thoracic CT imaging showing features highly suggestive of hypersensitivity pneumonitis, bronchoscopy with bronchoalveolar lavage showing CD8⁺ lymphocytosis, particularly if >50%, would provide additional sufficient evidence to establish the diagnosis of hypersensitivity pneumonitis for this patient.

Diagnosis: Hypersensitivity pneumonitis progressing to pulmonary fibrosis

References

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