Tuesday 15 October 2019

British Thoracic Imaging Society: Diagnosing usual interstitial pneumonia (UIP) in 2019 and why it is important: what the general radiologist needs to know

15:00 - 16:10

British Thoracic Imaging Society: Diagnosing usual interstitial pneumonia (UIP) in 2019 and why it is important: what the general radiologist needs to know

Dr Jonathan Rodrigues, Royal United Hospitals Bath NHS Foundation Trust

Learning points

Until recently, there were no effective treatments for idiopathic pulmonary fibrosis (IPF) and it had a very poor prognosis. However, two anti-fibrotic drugs (pirfenidone and nintedanib) have now been shown in randomised controlled trials to reduce disease progression compared to placebo.

CT is vital in the diagnosis of IPF. CT findings of 'typical usual interstitial pneumonia (UIP)' are a basal and subpleural predominant distribution with honeycombing, reticulation and traction bronchiectasis/bronchiolectasis and absence of features of alternative diagnoses. A 'typical UIP' pattern, in the correct context, allows a clinic-radiological diagnosis of IPF to be made in the MDT setting, allowing consideration of anti-fibrotic treatments.

The historic 'possible UIP' category on CT should now be subdivided into: 1) 'probably UIP' (as for 'typical UIP' but without honeycombing) and 2) 'indeterminate for UIP'. Whether 'probably UIP' on CT is sufficient for a clinic-radiological diagnosis of IPF or whether a surgical lung biopsy should be performed is a source of on-going debate.

CT features 'inconsistent with a UIP diagnosis' include upper or mid lung predominant distribution, peribronchovascular or perilymphatic predominance and subpleural sparing, predominant consolidation, extensive pure ground glass opacity (without acute exacerbation), extensive mosaic attenuation with extensive sharply defined lobular air trapping on expiration, diffuse nodules or cysts.

Ancillary CT features such as pleural plaques, dilated oesophagus, distal clavicular erosions, extensive lymph node enlargement, pleural effusions and pleural thickening suggest an alternative diagnosis to idiopathic pulmonary fibrosis, even in the presence of a 'typical UIP' or 'probable UIP' pattern.

References


British Thoracic Imaging Society: Diagnosing usual interstitial pneumonia (UIP) in 2019 and why it is important: what the general radiologist needs to know

Dr Graham Robinson, Royal United Hospitals Bath NHS Foundation Trust

Learning points

Differentiation between definite, probable and indeterminate features of UIP.

Definite:

- About 50% of cases\(^1\)
- Over 60 years, smokers, males
- Honeycombing with traction bronchiectasis and bronchiolectasis which is peripheral and subpleural – in non-specific interstitial pneumonia (NSIP) traction is often more central\(^2\)
- Ground glass with a fine reticular pattern and/or intrinsic traction bronchiectasis (not pure ground glass)
- Cranio-caudal gradient – basal with some upper lobe involvement
- Can be uniform cranio-caudally but need other features – otherwise consider indeterminate. 25% asymmetrical.
- Pulmonary ossification\(^3\)
- Some features of PPFE\(^4\)
- Consider coronals, sagittals and/or minimum UIP for bronchiectasis versus honeycombing
- Problems include paraseptal emphysema (especially at bases) versus honeycombing
- Reticulation is irregularly spaced with varying thin and thick lines cf NSIP where lines are often similar thickness and evenly spaced\(^2\)
- Subpleural sparing ‘absent’\(^5\)
- ‘Some’ areas of decreased lung attenuation allowable and do not necessarily point to chronic hypersensitivity pneumonitis (CHP)
- Spatial and temporal heterogeneity of NSIP spatial and temporal homogeneity
More diffuse cranio-caudal gradient or more upper lobe involvement seen in familial idiopathic pulmonary fibrosis (IPF)\(^6,7\)

**Probable\(^2\)**

- About 25-30\(^1\%

- As (above) definite UIP with basal, peripheral predominant fibrosis, traction bronchiectasis but no honeycombing or features of alternative diagnosis

- Can be diagnosed in diffuse zonal distribution if other findings of UIP present\(^8\)

**Indeterminate\(^2\)**

- About 25-30%

- Often driven by more diffuse axial and zonal distribution including peri-bronchovascular distribution and upper lobe involvement\(^9\)

- May just see non-specific reticulation but can have traction bronchiectasis

- ‘Some’ inconspicuous features of a non-UIP pattern

- Possible CTD related honeycombing\(^10\)
  - Anterior upper lobe sign/four-corner sign
  - Straight edge sign
  - Exuberant honeycomb sign
  - Pleural/pericardial effusion, oesophageal dilatation.