Mimics of chronic thrombo-embolic pulmonary hypertension (CTEPH)

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Learning objectives

- CTEPH is a potentially curable cause of pulmonary hypertension, yet the prognosis is very poor in untreated cases - accurate diagnosis is therefore vital.
- Many of the radiological features of CTEPH are non-specific and occur in other conditions.
- Distinguishing CTEPH from its mimics can be a radiological challenge.
- Awareness of the discriminating features of these conditions permits the correct radiological diagnosis to be made with more confidence.

Background

CTEPH results from progressive occlusion of the pulmonary arterial tree by thrombus following a single or recurrent acute pulmonary thromboembolism (PTE). Chronic thrombi interact with the pulmonary arterial endothelium, resulting in fibrotic deposits, with a secondary small vessel arteriopathy deep in the pulmonary vascular bed. Untreated, this condition carries a dismal prognosis. However, although surgical management still incurs significant morbidity and mortality, pulmonary thromboarterectomy in selected patients can offer excellent outcomes. Accurate diagnosis is therefore vital.

There are a number of characteristic radiological signs associated with CTEPH. However, many of these features are non-specific and are also demonstrated in other conditions which mimic CTEPH - namely pulmonary artery sarcoma, mediastinitis, vasculitis and Eisenmenger syndrome with giant pulmonary arteries and thrombosis in situ.

Whilst overlap of radiological features exists, awareness of the constellation of findings most characteristic of these respective conditions will enable radiological diagnoses to be made with increased confidence.

This presentation highlights the key radiological features which help to discriminate CTEPH from its mimics.

Imaging findings OR Procedure details

CTEPH
The following radiological features are characteristic of CTEPH, but are non-specific:\(^1\):

(1) Significantly enlarged pulmonary arteries, often asymmetrical
(Fig. 1).

(2) Multifocal perfusion : ventilation mismatch (Fig. 2).

(3) Central pulmonary artery thrombus, bronchial artery hypertrophy and mosaicism (Fig. 3, 4 & 5).

**Pulmonary artery sarcoma**

Pulmonary artery sarcoma is a rare malignancy arising from mesenchymal cells of the intima of the central pulmonary arteries. Its slow growth results in the insidious onset of chest pain, dyspnoea and cough.\(^2\)

The key discriminating radiological features are:

(1) Complete vessel occlusion (Fig. 6).

(2) Extraluminal extension (Fig 6).

(3) Vessel expansion (Fig. 7).

(4) Metastases (Fig. 7).

**Mediastinitis**

Mediastinitis is an uncommon condition characterised by the proliferation of fibrous tissue within the mediastinum and around the hila. The aetiology is usually sarcoidosis or other granulomatous diseases.

The key discriminating radiological features are:

(1) Central pulmonary artery stenosis (Fig. 8)

(2) Pulmonary vein involvement (Fig. 9)
(3) Airway obstruction (Fig. 10)

(4) Granulomatous disease e.g. mediastinal calcification, lymphadenopathy.

**Eisenmenger Syndrome with thrombosis in situ**

Prolonged increased flow through the pulmonary circulation in the context of congenital heart disease and a left to right shunt, can result in pulmonary hypertension and ultimately lead to flow reversal (Eisenmenger syndrome). Gross enlargement of the central pulmonary arteries is a characteristic feature and may be associated with thrombosis in situ.

The key discriminating signs are:

1. Clinical / radiological evidence of congenital heart disease and reversal of a left to right shunt.
2. Gross enlargement of the central pulmonary arteries, disproportionate in size to the rest of the pulmonary arterial tree, with central pulmonary artery thrombus (Fig. 11).
3. **Absence** of other vascular signs, such as vascular webs, vessel beading or paucity of peripheral arteries, beyond the central pulmonary arteries (Fig 12 & 13).

**Large vessel vasculitis**

This is arguably the most difficult condition to distinguish from CTEPH. Clinico-radiological correlation is particularly valuable in maximising the accuracy of radiological image interpretation. Clinical features such as fever, visual disturbance, headaches and claudication should raise suspicion of vasculitis. Evidence of elevated erythrocyte sedimentation rate and anaemia are also important.

Bronchial artery hypertrophy, mosaicism and a high probability VQ scan may occur in this condition. However, these findings are considerably more common in CTEPH. Disease confined to the large central pulmonary arteries is also a salient feature of vasculitis, but a similar distribution of disease is seen in thrombosis in situ.

The discriminating radiological features are:
(1) Abnormal aortic contour and mural thickening (Fig. 14)

(2) Aneurysms

**Summary**

A summary table of the discriminating features of each condition.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Discriminating features</th>
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| CTEPH                                               | • Intraluminal thrombus throughout the pulmonary arterial tree  
                                                       • Vascular webs, beading and pruning of peripheral pulmonary arteries  
                                                       • Increased incidence of bronchial artery hypertrophy, mosaicism and high probability VQ scan compared with the "mimics" |
| Pulmonary Artery Sarcoma                            | • Large volume filling defects in PA with total/sub-total vessel occlusion  
                                                       • Extraluminal extension  
                                                       • Metastases |
| Mediastinitis                                       | • Central PAstenosis  
                                                       • Pulmonary vein involvement  
                                                       • Airway obstruction  
                                                       • Granulomatous mediastinal disease |
| Eisenmenger syndrome with thrombosis in situ        | • Congenital heart disease with reversal of a left to right shunt  
                                                       • Gross central pulmonary artery enlargement |
• Absence of peripheral vascular abnormalities
• Abnormal aortic contour
• Aneurysms
• Arterial calcification
• Absent intra-luminal thrombus

Images for this section:

Fig. 1: Plain chest radiograph demonstrating asymmetrical significant enlargement of the pulmonary arteries (arrow) in a patient with CTEPH.
Fig. 2: VQ scan demonstrating multiple regions of ventilation : perfusion mismatch in a patient with CTEPH.
Fig. 3: CT pulmonary angiogram demonstrating central pulmonary artery thrombus (asterisk) in a patient with CTEPH.
Fig. 4: Bronchial artery hypertrophy (arrows), in a patient with CTEPH.
**Fig. 5:** Mosaicism and enlargement of the segmental pulmonary arteries relative to their accompanying bronchi, in a patient with CTEPH.
Fig. 6: CT pulmonary angiogram in a patient with pulmonary artery sarcoma. Note the large intraluminal filling defect in the central pulmonary arteries (asterisks). Although this feature is shared with CTEPH, filling defects causing complete or sub-total occlusion are more suggestive of pulmonary artery sarcoma. Extraluminal extension of tumour is also demonstrated (arrow).
**Fig. 7:** Contrast enhanced CT in a patient with pulmonary artery sarcoma. Intrapulmonary metastases are demonstrated in the right lung (arrows). In addition, large filling defects within the visualized pulmonary arteries are demonstrated bilaterally, with vessel expansion (asterisks).
Fig. 8: CT pulmonary angiogram in a patient with mediastinitis. Stenosis of the right interlobar artery is secondary to soft tissue encasement (arrow). Recanalisation of the interlobar artery in CTEPH can produce a similar appearance.
**Fig. 9:** Coronal maximum intensity projection reconstruction of a CT pulmonary angiogram demonstrating irregularity and encasement of the left superior pulmonary veins (arrow) in a patient with mediastinitis.
Fig. 10: CT pulmonary angiogram on lung window settings in a patient with mediastinitis. Note the bronchial stenosis (arrow) and the enlarged central pulmonary arteries (asterisk).
**Fig. 11:** CT pulmonary angiogram in a patient with Eisenmenger syndrome and thrombosis in situ. Note the disproportionate enlargement of the central pulmonary arteries (arrow) and the large eccentric thrombus, which is confined to the central pulmonary arteries (asterisk).
**Fig. 12:** Coronal maximum intensity projection of a CT pulmonary angiogram in a patient with CTEPH. Note the vascular web within the left pulmonary artery (arrow) - the absence of this feature is supportive of thrombosis in situ.
Fig. 13: Conventional pulmonary angiogram in a patient with CTEPH. Note the paucity of peripheral sub-segmental vessels (asterisks). The absence of this finding is supportive of thrombosis in situ.
**Fig. 14:** CT pulmonary angiogram in a patient with vasculitis, demonstrating mural thickening and an abnormal contour of the main and left pulmonary arteries (asterisk). This appearance was originally reported as pulmonary artery thrombus secondary to CTEPH. The patient underwent thoracotomy - surgical findings suggested an alternative diagnosis, thus highlighting the significant diagnostic challenge this condition presents.
Conclusion

CTEPH is unique amongst the causes of pulmonary hypertension due to the existence of effective surgical treatment. Accurate diagnosis is therefore vital. There are several characteristic radiological features of CTEPH but they are non specific and shared with other conditions. This overlap can hamper accurate diagnosis, but awareness of the key discriminating imaging features in each of these conditions increases confidence in radiological assessment and diagnosis.

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References

