Multi modality assessment of the pulmonary arteries: Distinguishing pulmonary artery sarcoma from chronic thromboembolic pulmonary hypertension

Poster No.: P-0119
Congress: ESTI 2015
Type: Educational Poster
Authors: N. F. Bassett, J. Tanner, S. Karia, K. Tweed, N. J. Screaton; Cambridge/UK
Keywords: Neoplasia, Education, PET-CT, MR, CT-Angiography, Thorax, Pulmonary vessels, Cardiovascular system
DOI: 10.1594/esti2015/P-0119

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.
As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method ist strictly prohibited.
You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.
Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.
www.myESR.org
Learning objectives

- Describe the typical imaging appearances of CTEPH using a multimodality approach at diagnosis and following treatment of pulmonary endarterectomy.
- Demonstrate appearances of pulmonary artery sarcoma (PAS) using multimodality imaging.
- Delineate the key imaging features to differentiate CTEPH from pulmonary artery sarcoma.
- Illustrate mimics of PAS, including pulmonary valve endocarditis.
Background

Pulmonary artery sarcoma

PAS was originally described by Mandelstamm in 1923 and was enough of a rarity for some contemporary authors to refute its existence. These tumours tend to arise from the intima and macroscopically appear as gelatinous clots filling the lumen of the artery. Most involve both pulmonary arteries (85%) and the pulmonary trunk. Less involve the pulmonary valve itself (35%). Patients usually present with pulmonary artery obstruction, pulmonary hypertension and right ventricular failure.¹²³

PAS is thought to arise predominantly from pluripotent intimal stem cells and the histological diagnosis is usually that of undifferentiated intimal sarcoma or leiomyosarcoma.⁴⁵

Unfortunately the prognosis of PAS is poor, largely due to the late presentation as the clinical symptoms do not normally occur until pulmonary vessels become completely occluded. The diagnosis is also delayed due to the non-specific clinical picture and imaging overlap with other conditions (such as pulmonary hypertension, heart failure and CTEPH). The treatment of choice is surgical resection by endarterectomy and chemotherapy and the average survival is 1-2 years.⁶

Chronic thromboembolic pulmonary hypertension

CTEPH is a rare and severe condition which is characterised by the presence of single or recurrent pulmonary thromboemboli that fail to resolve. Recent research suggests that this condition is not caused purely by pulmonary vascular obliteration of elastic arteries from the unresolved pulmonary emboli (the underlying reason for non-resolution of these emboli is not yet known). Persistent obstruction of pulmonary arteries by organised thrombus leads to elevated pulmonary artery pressures and increased shear stresses in pulmonary vessels not directly occluded by thromboembolism. This in turn results in small vessel pulmonary vascular disease re-modelling identical to that seen in pulmonary artery hypertension with the clinical outcome of progressive pulmonary hypertension. Thus the proximal pulmonary artery occlusion and secondary small vessel disease contribute to the elevated pulmonary vascular resistance. The severity of the secondary small vessel disease also correlates with the risk of surgery and post-surgical outcome.⁷
CTEPH often presents with progressive dyspnoea, haemoptysis and signs of right heart failure (fatigue, palpitations, syncope, or oedema) after single or recurrent pulmonary emboli. There is usually an interval time period of months to years between the initial event and the development of clinical symptoms.\(^7\)

Low velocity flow blood in distal vessels can also lead to peripheral in-situ thromboses (this occurs in approximately 20\% of cases), which are not amenable to pulmonary endarterectomy but the associated vasculopathy may be amenable to targeted medical therapy. Pulmonary endarterectomy (PTE) therefore remains the preferred treatment in order to maximally reduce the pulmonary arterial pressures but is only suitable if there is a proximal distribution of disease. PTE has a documented 6 year survival rate of approximately 75\%.\(^8,^9\) Balloon pulmonary angioplasty is currently being evaluated in patients considered inoperable due to co-morbidity or disease distribution or in patients with persistent pulmonary hypertension following pulmonary endarterectomy.
Imaging findings OR Procedure details

Although plain chest radiographs are most often performed as first line investigations, the findings are usually non-specific (possibly showing evidence of pulmonary hypertension or evidence of regional oligoemia/pulmonary infarcts). Similarly, ventilatory/perfusion scintigraphy provides similar imaging findings between CTEPH and PAS and cannot give direct visualisation of the filling defects themselves. Echocardiography is frequently used in the work up of pulmonary hypertension, however this is performed primarily to examine real time valvular and cardiac function rather than for visualisation of the pulmonary arterial defects. The distinguishing features of CTEPH and PAS are currently best demonstrated on CT, MRI or PET/CT imaging.

CT pulmonary angiography is frequently the first investigation that allows possible differentiation between CTEPH and PAS based on the morphology of the filling defects and the distribution of findings. CT also provides high spatial resolution detail of the thoracic anatomy. The primary disadvantage of CT is the radiation burden, however it is a fast and readily available test.

MRI gives further information with improved tissue characterisation and enhancement pattern with administration of intravenous gadolinium. Details of the cardiac valvular and ventricular function may also be obtained. The main disadvantages of MRI are the prolonged length of scan time, poor spatial resolution and high specialisation of this technique which would make it less available in comparison to CT.

PET/CT can also be used to differentiate between CTEPH and PAS due to the increased FDG tracer uptake of PAS in comparison to thrombus. This technique is also useful for demonstrating metastatic deposits.

Chronic thromboembolic pulmonary hypertension imaging findings

*CT findings:* CT often demonstrates multiple bilateral filling defects with distal vessel involvement and mosaic attenuation.

Pulmonary arterial filling defects (Figs 1&2):

- Multiple hypoattenuating bilateral filling defects within the pulmonary arteries on CT pulmonary angiography
- Distal vessel involvement with vessel attenuation/truncation
- Webs and bands
- Calcification of thrombus
Evidence of pulmonary hypertension (Fig 3):

- Right heart strain with dilatation of the right sided cardiac chambers and reflux of contrast medium into the IVC and hepatic veins in keeping with tricuspid regurgitation
- Dilatation of the main pulmonary artery greater than or equal to 29mm
- Right ventricular hypertrophy, dilatation and trabeculation

Mosaic attenuation is often demonstrated in the lung parenchyma due to oligaemia around occluded pulmonary arteries causing regions of hypoattenuating lung parenchyma and decreased pulmonary vessel size within these areas (Fig 3)

MRI findings: As with CT, the filling defects are again demonstrated on MRI however there is also the additional information provided by perfusion imaging and evaluation of cardiac function.

Pulmonary arterial filling defects

Parenchymal perfusion defects on perfusion imaging (Fig 2)

Further evidence of pulmonary hypertension\textsuperscript{10}:

- Reduced peak velocity flow in the pulmonary arteries and increased retrograde flow during post-systolic phase
- Right ventricular hypokinesis
- Decreased ejection fraction and cardiac output

PET/CT findings: PET/CT is not commonly used in the evaluation of CTEPH, however the following features have been documented previously.

- None to low grade FDG uptake
- A small study suggested a correlation between increased FDG uptake in the right ventricular myocardium correlated and the severity of right ventricular overload\textsuperscript{11}

Pulmonary artery sarcoma imaging findings

CT findings: Unlike the filling defects in CTEPH, PAS is more often suspected with unilateral and contiguous proximal disease. Extension beyond the vessel and metastatic spread are also indicators of PAS.

Pulmonary arterial filling defects (Fig 4):

- Single hypoattenuating filling defect on CTPA
• Originating in a proximal pulmonary artery
• Contiguous filling defect
• Causes complete or sub-total occlusion
• Convex medial margin
• Expands the pulmonary artery (Fig 5)
• Thickened vessel walls where there is tumour involvement (Fig 6)
• May demonstrate delayed contrast enhancement

Evidence of pulmonary hypertension (Fig 7):

• Same findings as described under CTEPH

Evidence of metastatic spread with pulmonary/mediastinal/bony metastases

MRI findings: On MRI the arterial filling defects have the same morphology as described on CT, however the addition of gadolinium allows better distinction between tumour and thrombus.

• Filling defects in the pulmonary arteries (Fig 6)
• Enhances with administration of Gadolinium
• Allows tumour to be distinguished from coexistent thrombus

Further findings of pulmonary hypertension:

• Same as findings described under CTEPH

PET/CT findings:

• PAS demonstrates increased FDG uptake (Fig 6).

Imaging findings of other possible PAS mimics

Thrombosis in situ with Eisenmenger syndrome (Fig 8)

• CT demonstrates the right to left shunt as well as dilated central pulmonary arteries with eccentric pulmonary artery thrombus

Large vessel vasculitis

• CT demonstrates circumferential pulmonary arterial intimal thickening in affected regions
• MRI also demonstrates intimal thickening of the affected pulmonary arteries
• PET/CT shows intermediate FDG uptake in the region of the vasculitis

Pulmonary valve endocarditis
• CT demonstrates a filling defect in the region of the pulmonary valve which does not extend beyond the vessel
• PET/CT shows increased FDG uptake in the region of inflammation

**Differentiation between PAS and CTEPH**

While endarterectomy is commonly the primary therapeutic modality for both it is important that the surgeon is aware of the likely diagnosis of PAS as operative approach differs and features of non-resectability such as extra-luminal invasion and metastases should be actively sought. Particular features which should lead to PAS being considered are purely unilateral disease involving the main pulmonary artery/pulmonary trunk with luminal expansion and a convex medial margin. (See Fig 9 for a further breakdown of the distinguishing features)
Fig. 1

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK
Fig. 2

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK

Fig 2. CTEPH in the same patient demonstrating:
A) Multiple non occlusive filling defects in the right upper lobe pulmonary artery
B) Larger filling defect occluding the right main pulmonary artery as well as a non occlusive thrombus in the left lower lobe pulmonary artery
C) Distal vessel attenuation in the right lower lobe and distal thrombus in a segmental branch of the left lower lobe
D) MRI perfusion scan demonstrating reduction in the right lower lobe perfusion

Fig 3. CTPA of CTEPH in the same patient demonstrating:
A) Dilated MPA
B) Small distal sub-segmental emboli
C) Dilated right heart chambers
D) Mosaic attenuation
E) Tricuspid regurgitation
Fig. 3

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK

Fig 4. PAS occluding the right pulmonary artery with paucity of distal arterial opacification

Fig. 4

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK
Fig. 5

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK

Fig 5. Comparison of PAS on MRI and CTPA demonstrating diffuse infiltrating morphology of the tumour expanding the left pulmonary artery.
Fig. 6

A) CTPA demonstrating a contiguous unilateral right main pulmonary artery filling defect

B) The same findings on MRI

C) PET study demonstrating increased FDG uptake within the right main pulmonary artery lesion
Fig. 7

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK
Fig. 8

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK
<table>
<thead>
<tr>
<th>Distinguishing Features</th>
<th>PAS</th>
<th>CTEPH</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CTPA</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery filling defects:</td>
<td>Proximal</td>
<td>Pulmonary artery filling defects:</td>
</tr>
<tr>
<td></td>
<td>Contiguous</td>
<td>- Non-occlusive</td>
</tr>
<tr>
<td></td>
<td>Complete/Sub-total occlusion</td>
<td>- Multiple</td>
</tr>
<tr>
<td></td>
<td>Distends the vessel</td>
<td>- Bilateral</td>
</tr>
<tr>
<td></td>
<td>Convex medial margin</td>
<td>- Webs/Bands</td>
</tr>
<tr>
<td></td>
<td>Extension beyond the vessel</td>
<td>Mosaic attenuation</td>
</tr>
<tr>
<td></td>
<td>Possible delayed contrast enhancement</td>
<td></td>
</tr>
<tr>
<td>Metastases</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>MRI</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Enhancement with gadolinium</td>
<td></td>
<td>No enhancement</td>
</tr>
<tr>
<td><strong>PET/CT</strong></td>
<td>High FDG uptake</td>
<td>None or low FDG uptake</td>
</tr>
</tbody>
</table>

**Fig. 9**

© Radiology, Papworth Hospital NHS Trust, Papworth Hospital NHS Trust - Cambridge/UK
Conclusion

Although PAS is a rare diagnosis, it is essential that the reporting radiologist is aware of the imaging findings in order to raise it as a possibility when considering non-resolving filling defects within the pulmonary arteries.

As described previously, both PAS and CTEPH can manifest with severe pulmonary hypertension as well as similar clinical symptoms. Currently a combined analysis of CT, MRI and/or PET/CT imaging would allow for more confidence when differentiating between PAS and CTEPH. Understanding the key imaging appearances using a multimodality approach with comparison of the discriminating features is fundamental for appropriate diagnosis and treatment.
References

Personal Information

Nicholas Bassett BSc MBBS FRCR

Radiology Registrar

Papworth Hospital NHS Foundation Trust

Papworth Everard

Cambridge

UK

CB23 3RE

Telephone: 01480 830541

Email: NicholasBassett@nhs.net