

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ
الْحَمْدُ لِلَّهِ الَّذِي
خَلَقَ السَّمَوَاتِ وَالْأَرْضَ
وَالَّذِي يُضَوِّبُ الْمَوْتِ
الَّذِينَ آمَنُوا وَأَتَتْ
رَبَّهُمْ بِالْإِيمَانِ وَالْجَمِيلِ

Congenital & Acquired Pulmonary Artery Anomalies in the Adult

Dr. Mohamad Hasan Alkousy

Ass. Professor of Radiology

Sohag University

- In normal adults, the pulmonary trunk (main P.A.) may have a diameter as great as 28 mm.
- The main, Lt & Rt PAs are intrapericardial.
- The Rt P.A. has a longer mediastinal course than the Lt and it divides into two lobar branches at the root of the Rt lung.
- The Lt P.A. courses over the Lt main bronchus and penetrates the root of the Lt lung, where the artery divides into two lobar branches.
- The Rt & Lt P.As. should be of approximately equal size, although the Lt appears slightly larger in most subjects.
- The segmental arteries are always seen near the accompanying branches of the bronchial tree.

When the presence of a P.A. anomaly is suspected on the basis of chest radiography, CT is invaluable for examining the pulmonary vessels.

Advantages of CT

- Can provide clear depiction of peripheral vessels.
- Has an advantage over angiography in that it allows the lung parenchyma and the heart to be evaluated at the same time as the vessels.

a-Congenital lesions

- 1-Proximal interruption.
- 2-Anomalous origin of Lt P.A. (P.A. sling).
- 3-Idiopathic dilatation of pulmonary trunk.

b-Acquired lesions

- 1-Dilatation
 - P.A. hypertension.
 - Aneurysm.
 - Intravascular metastasis.

- 2-Narrowing
 - Bronchogenic ca.
 - Mediastinal fibrosis
 - Takayasu arteritis.

- 3-Thromboembolism & P.A. sarcoma.

a)-Congenital Lesions

**1-Unilat. proximal interruption of P.A.
(Rt / Lt)**

- Uncommon developmental anomaly (Rt is common than Lt)
- Means absence of the P.A. (ends blindly at the hilum).
- The term (interruption) is used in preference to (absence) of a pulmonary artery, since the portion of the vessel that is in the lung is usually intact and patent.
- Blood is supplied to the lung via collateral systemic vessels, mainly bronchial arteries but also via transpleural branches.

-X-Ray → ↓Volume of hemithorax (diaphragmatic & mediastinal shift)

→ No air trapping on expiration.

→ Hyperinflation & herniation of other lung.

→ Later, pul. hypertension changes.

-CT → Abscent mediastinal part of affected P.A.

/→ May terminate within 1 cm of its origin.

→ Collaterals (Serrated thickening of the pleura and subpleural parenchymal bands).

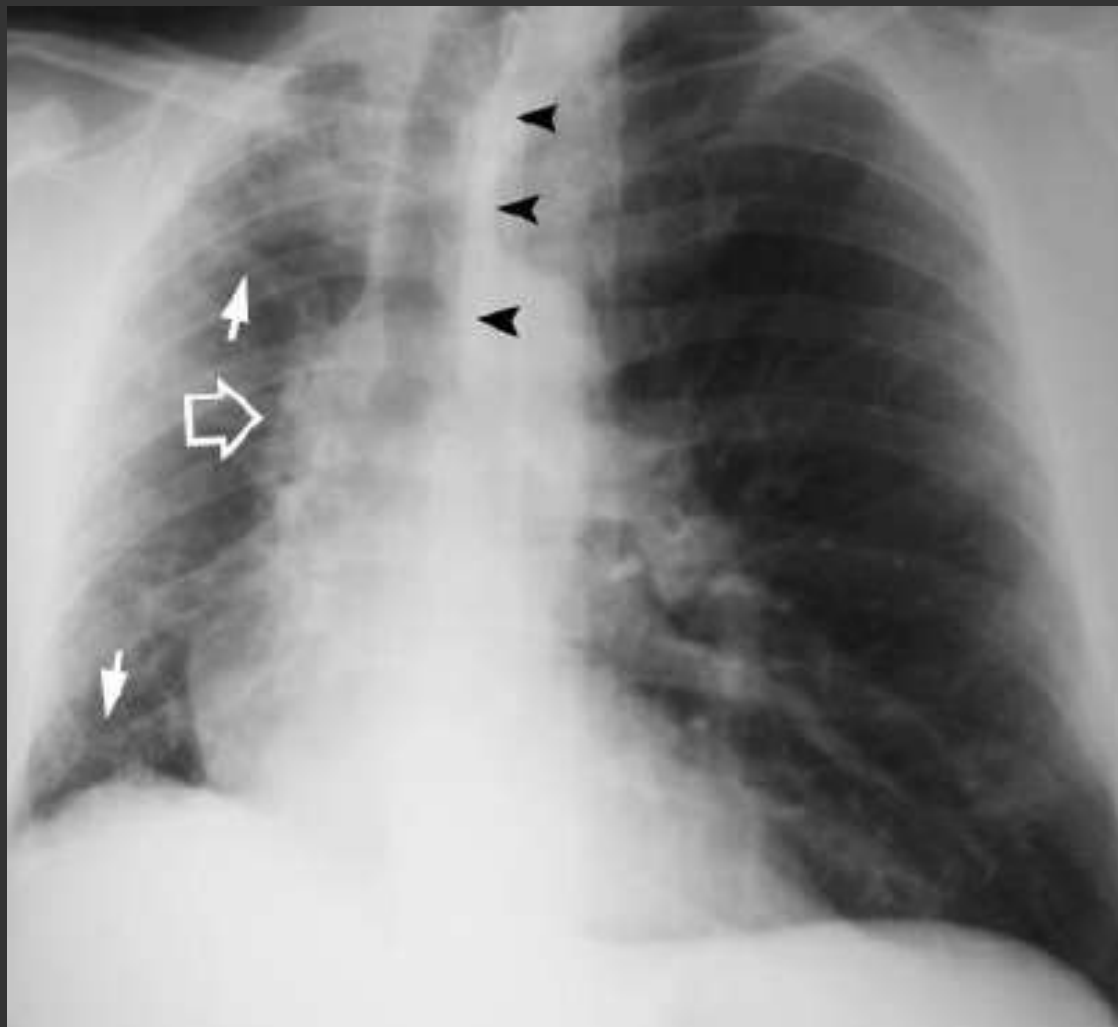


Fig 1. Asymptomatic unilat. proximal interruption of Rt P.A. in a 48 y old man.

There is small Rt hemithorax, mediastinal shift (arrowheads), absence of the Rt. P.A. shadow (open arrow), and linear opacities that correspond to systemic collateral vessels (solid arrows) along the pleura and within the lung.

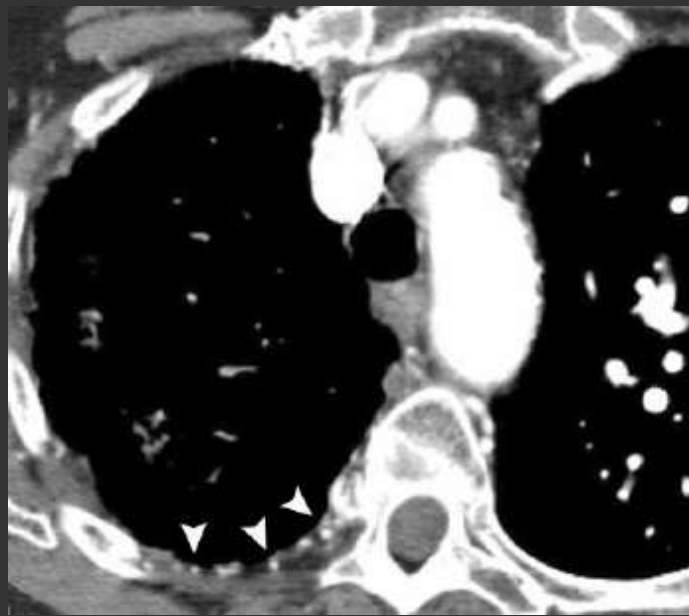


Fig 2. Unilat. proximal interruption of the Rt P.A. in a 52 y old woman with progressive shortness of breath and hemoptysis.

- (a) Enhanced CT shows only the proximal portion of Rt P.A. (arrowhead) and enlargement of main & Lt P.As. that indicates pulmonary hypertension.
- (b) Enhanced CT scan at the level of upper lobes shows serrated thickening of the Rt pleura because of enlarged intercostal collateral vessels (arrowheads).
- (c) Lung window shows multiple linear opacities perpendicular to the pleural surface that correspond to transpleural systemic vessels (arrowheads).

**2-Anomalous origin of Lt P.A.
(P.A. sling)**

-Rare vascular developmental anomaly.

-The Lt P.A. arises from the posterior aspect of Rt P.A. and passes between the trachea and esophagus to reach Lt hilum.

-The Lt P.A. thus forms a sling around the distal trachea and the proximal Rt main bronchus.

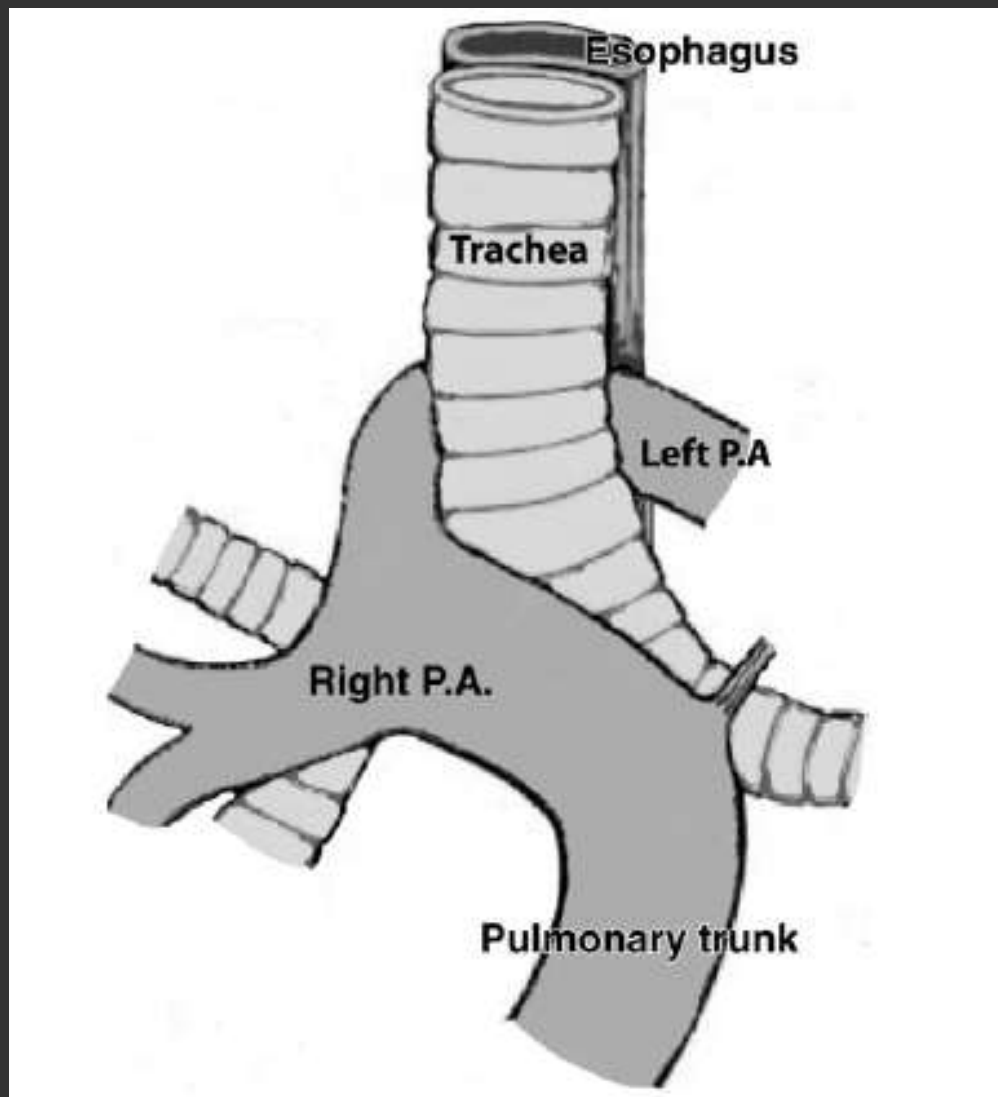


Fig 3. Diagram shows the anomalous origin of a Lt P.A. that arises from the posterior aspect of the Rt P.A. and reaches the Lt hilum by passing between the trachea and esophagus.

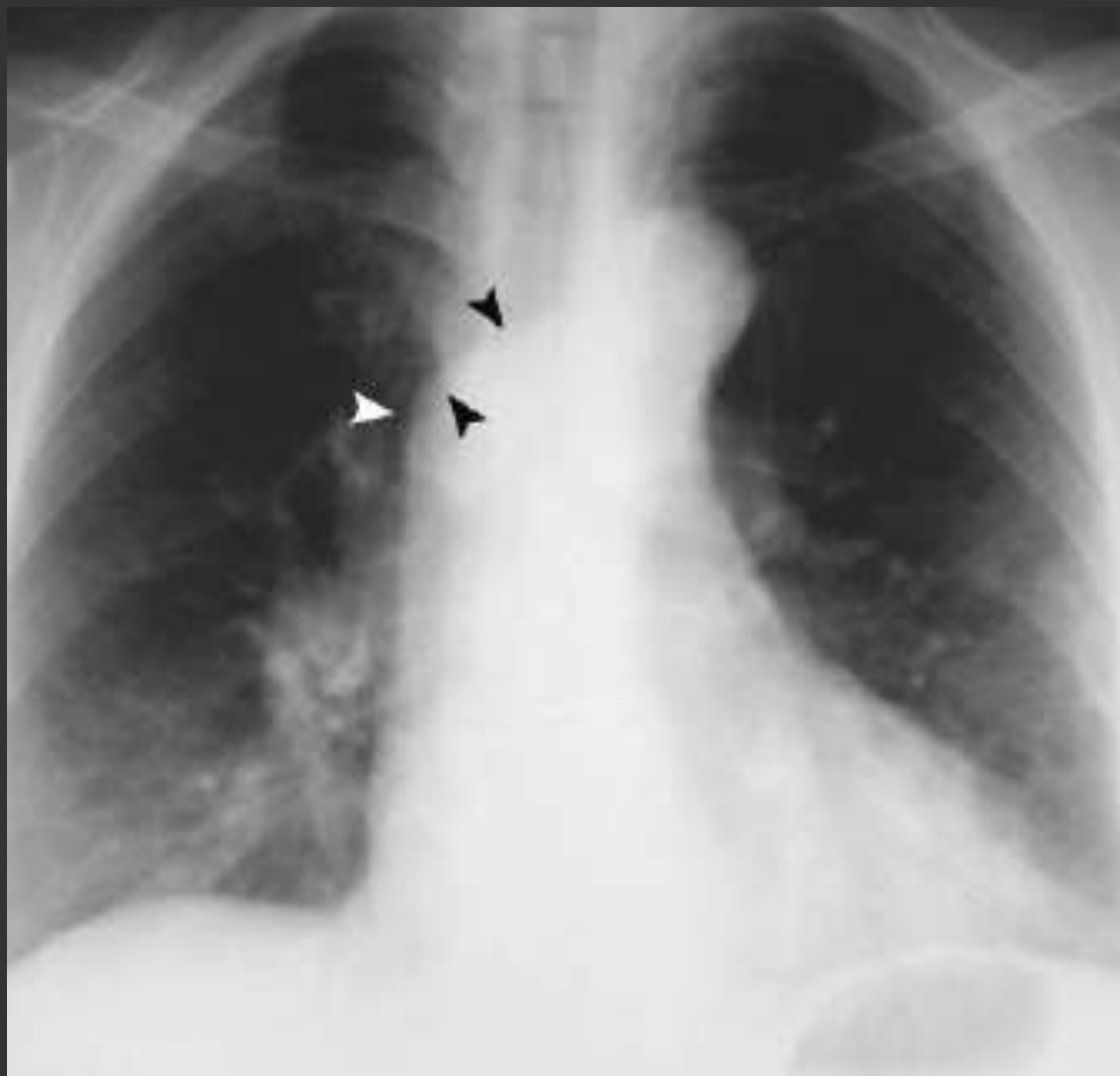


Fig 4. Anomalous origin of the Lt. P.A. in a 60 y old asymptomatic woman.

(a) Chest radiograph shows an anomalous Rt paratracheal border (arrowheads).

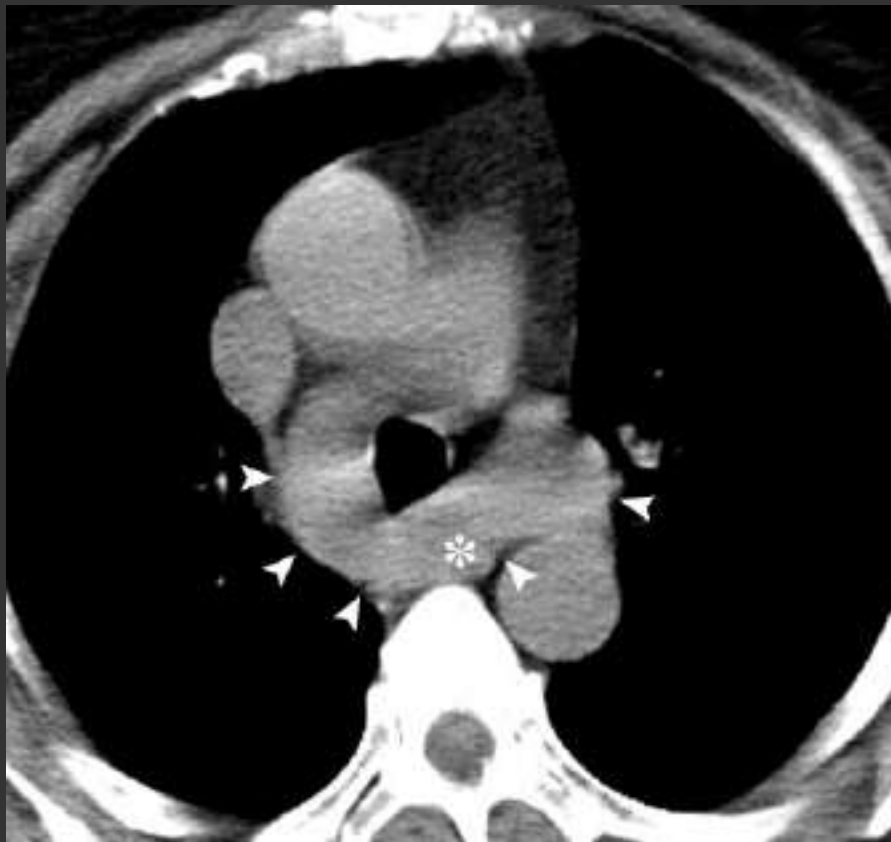


Fig 4. Anomalous origin of the Lt. P.A. in a 60 y old asymptomatic woman.

Unenhanced CT (**b**) and MRA (**c**) at the level of pulmonary trunk show the abnormal course of the Lt P.A. (arrowheads in **b**) between the lower portion of the trachea and esophagus (* in **b**).

3-Idiopathic dilatation of pulmonary trunk

-Rare congenital anomaly that involves abnormal enlargement of the pulmonary trunk.

-Occurs with / without dilatation of the Rt & Lt P.As.

-We must exclude pulmonary and cardiac diseases (pulmonary valve stenosis) and to confirm the presence of normal pressure in the Rt ventricle and P.A.

-Patients are asymptomatic and the anomaly is usually detected accidentally on chest radiographs or CT scans.

X-Ray → Enlarged main P.A. → rounded bulge at Lt mediastinal border (mimic mass).

Enhanced CT / MRI / Echo → confirm diagnosis.

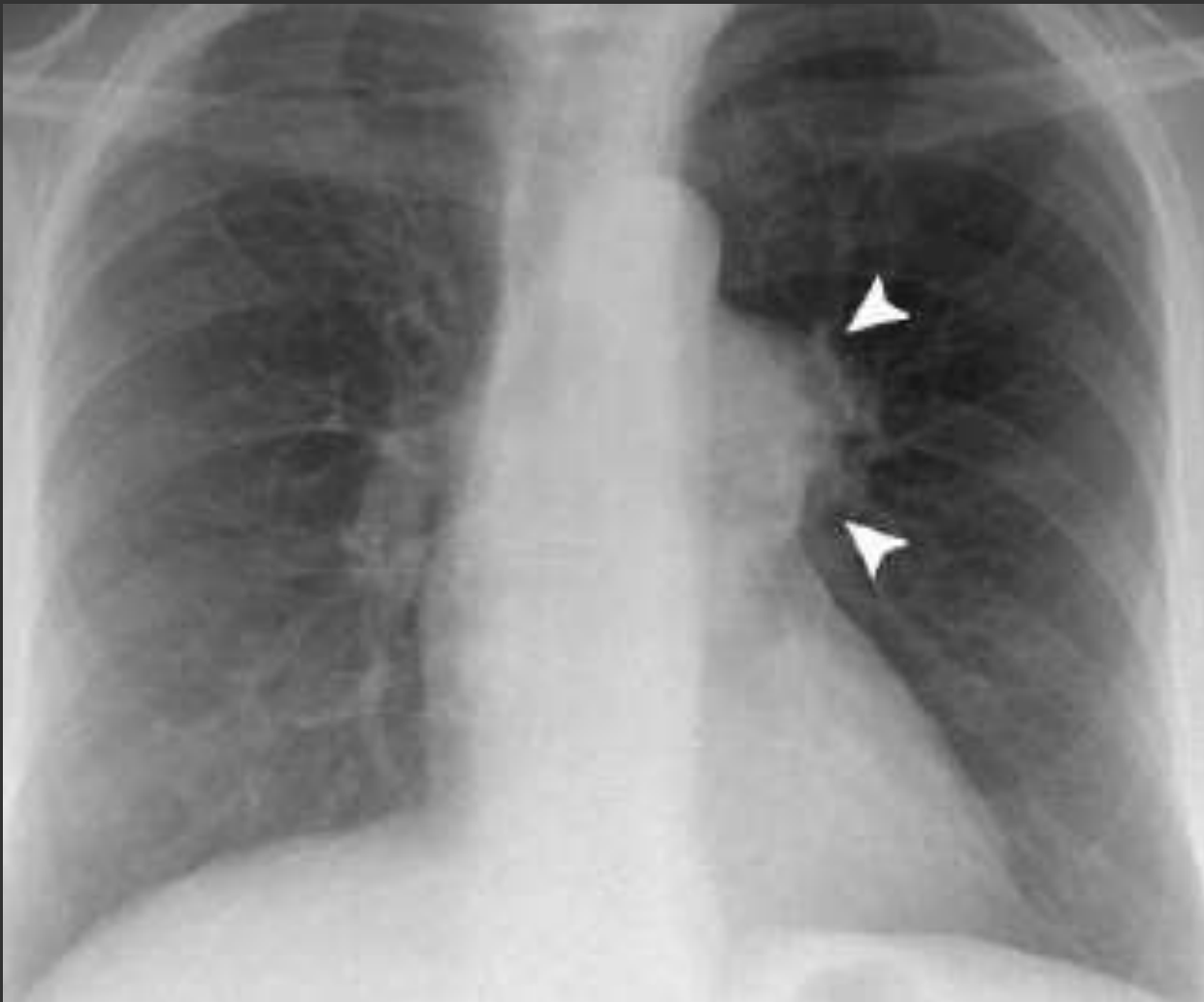


Fig 5. Idiopathic dilatation of the pulmonary trunk in a 55 y old asymptomatic woman.

(a) Chest radiograph shows an abnormal bulge in the Lt mediastinal border (arrowheads), a feature suggestive of a mediastinal mass.

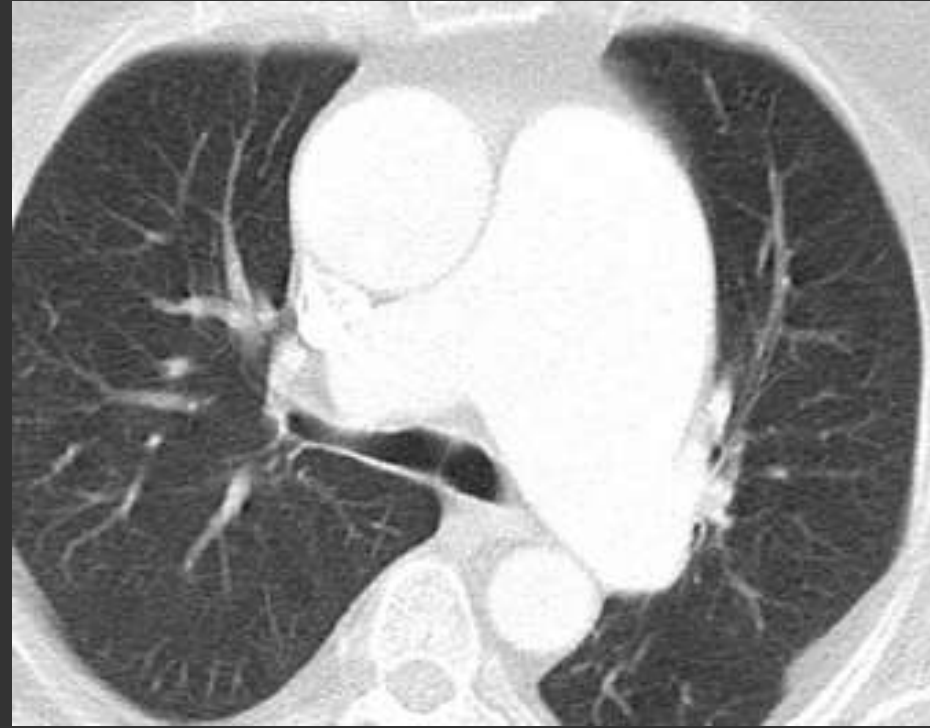


Fig 5. Idiopathic dilatation of the pulmonary trunk in a 55 y old asymptomatic woman.

(b) Enhanced CT shows abnormal enlargement of the main pulmonary trunk, with mild dilatation of the Rt & Lt P.As. **(c)** Lung window at same level as **b** shows normal vessels and parenchyma.

b)-Acquired Lesions

P.A. Dilatation

Include

1-P.A. hypertension.

2-P.A. Aneurysm.

3-Intravascular metastasis

1-Pulmonary Hypertension

-Pulmonary hypertension is the most frequent cause of diffuse enlargement of the pulmonary arteries.

-The 2ry type (cardiac, pulmonary or hepatic) is more common than 1ry type (idiopathic).

-Pulmonary hypertension means that P.A. pressure is

↑25 mm Hg at rest (normal level = 10 mm Hg)

or ↑30 mm Hg during exercise (normal level = 15 mm Hg).

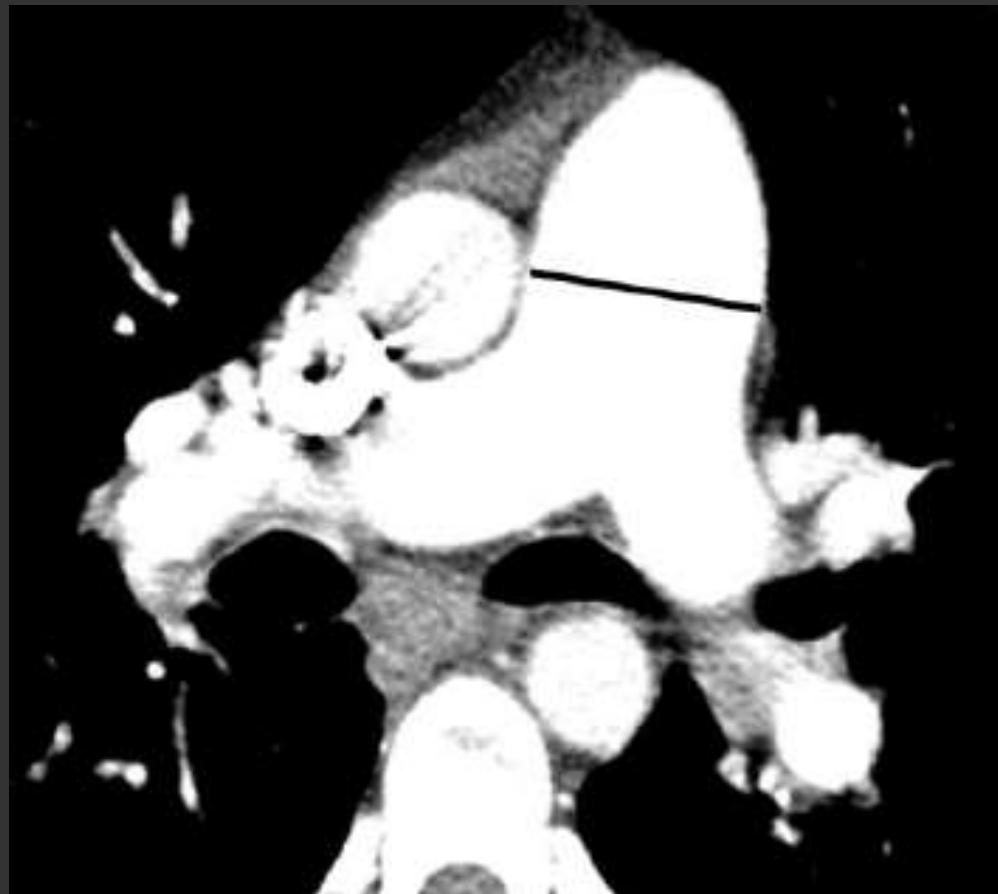
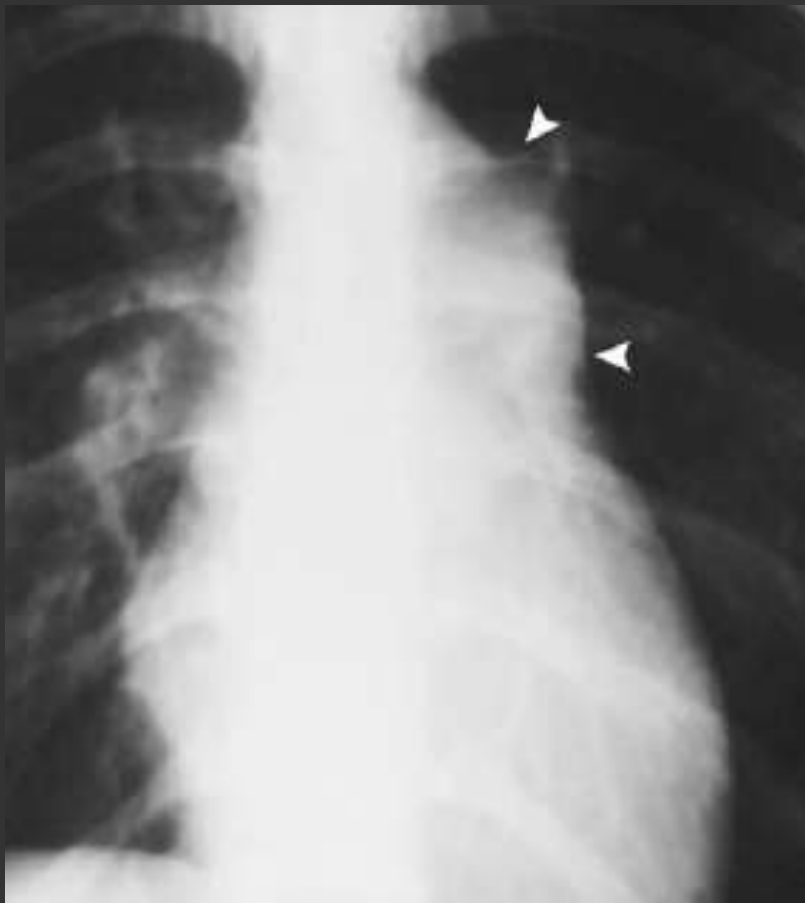


Fig 6. Pul. hypertension in a 32 y old woman with HIV infection.

(a) Chest radiograph shows enlargement of the main P.A. (arrowheads). **(b)** Enhanced CT shows an enlarged pulmonary trunk with a maximum diameter of 39 mm (black line) near its bifurcation, lateral to ascending aorta (a diameter greater than that of ascending aorta).

1-Vascular Signs

- Dilated main central P.A. (diameter ↑29 mm) (↑than aorta).
- Peripheral vasoconstriction.
- Dilated bronchial artery branches (++ with ch. thromboembolic pulmonary hypertension than with the 1ry type).
- Complications → thrombosis, P.A. calcific atheromas, dissecting aneurysm.

2-Lung parenchyma signs

-Mosaic pattern (patchy areas of ↓ / ↑ attenuation).

D.D. Airway anomalies

(enlarged MPA suggest pul. hypertension)

(dilated bronchi suggest airway abnormality).

3-Mediastinal & Cardiac signs

-Rt V. enlargement (Rt V : Lt V ratio is ↑ 1:1).

-Lt sided bowing of interventricular septum.

-↑Rt. V. myocardial thickness (↑ 4 mm).

-Dilated IVC.

-Pericardial thickening / small effusion.

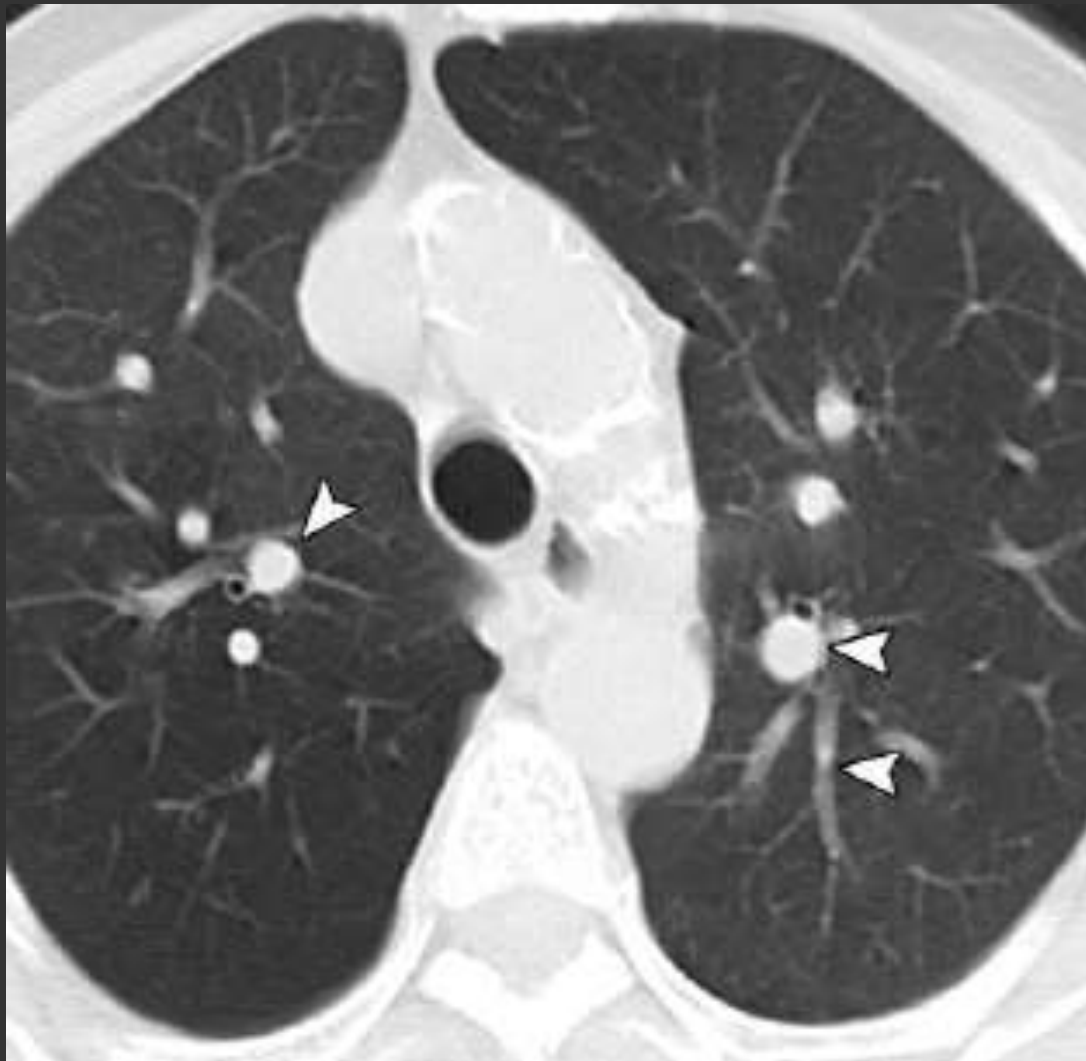


Fig 7. Pul. hypertension in a 75 y old man.

CT scan (lung window) at the level of upper lobes shows marked enlargement of the pulmonary arteries (arrowheads) in relation to the bronchi.



Fig 8. Pul. hypertension in a 65 y old woman.

CT scan (lung window) shows a mosaic perfusion pattern, with increased diameters of vessels in areas of hyper-attenuation (arrows) and sharp tapering of peripheral vessels in areas of hypo-attenuation (arrowheads).

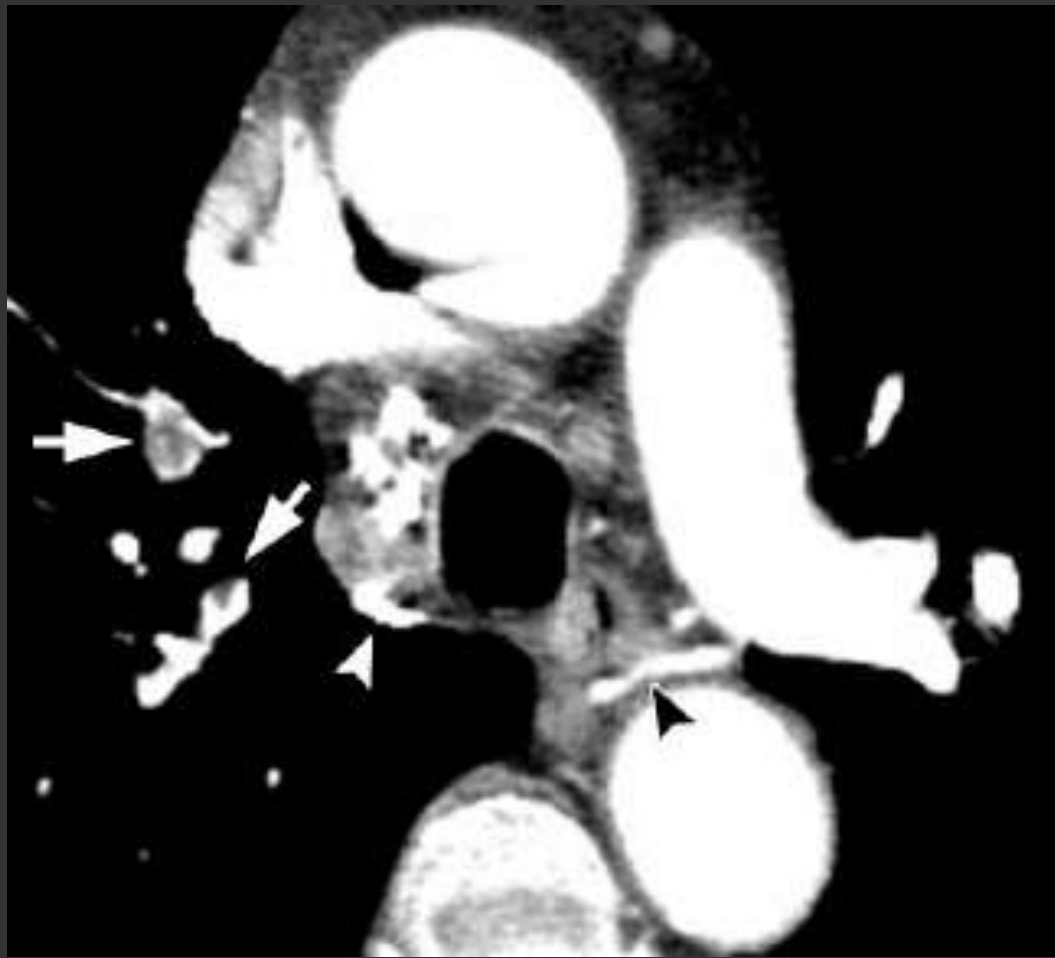


Fig 9. Ch. thromboembolic pul. hypertension in a 62 y old man with dyspnea.

Enhanced CT scan shows enlargement of Lt (black arrowhead) and Rt (white arrowhead) bronchial arteries, as well as filling defects in the Rt upper lobe pulmonary vessels (arrows) that correspond to new locations of acute pulmonary thromboembolism.

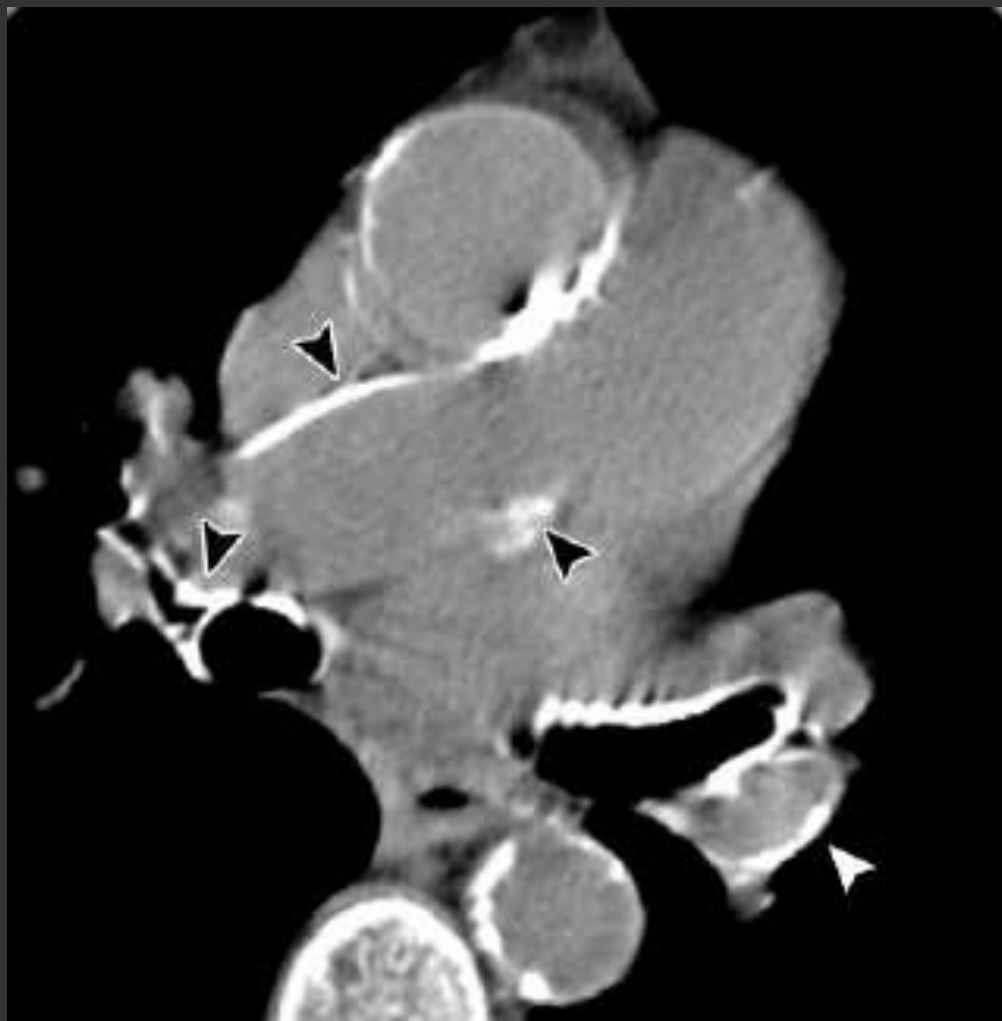


Fig 10. Pul. hypertension in a 72 y old man with a mitral valve abnormality.

Unenhanced CT scan shows dilatation and atherosclerotic calcification of the main & Rt P.As. (black arrowheads) and Lt interlobar artery (white arrowhead).

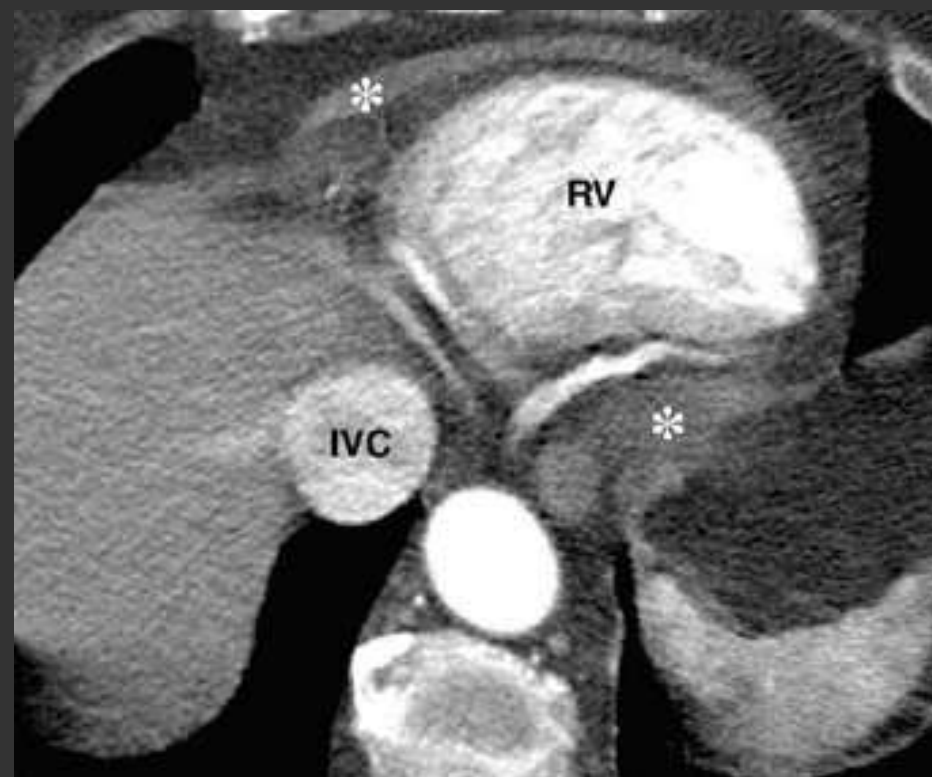
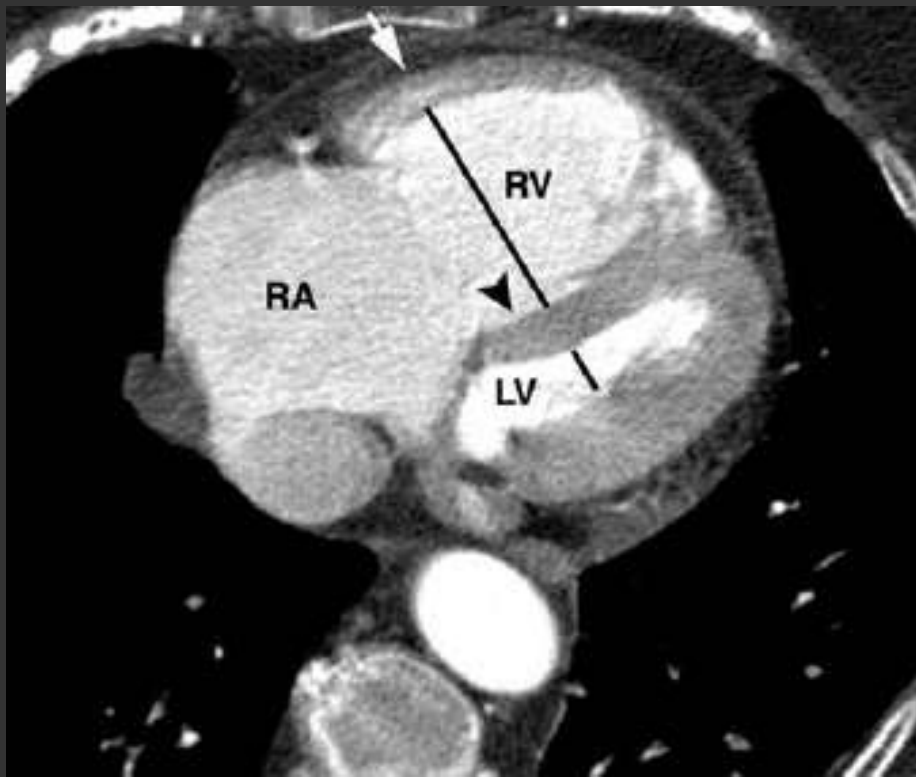


Fig 11. Rt heart abnormalities 2ry to pul. hypertension in a 56 y old woman.

(a) Enhanced CT scan shows dilatation of the Rt. ventricle (*RV*), with a Rt V./ Lt V. (*LV*) ratio greater than 1:1; Lt sided septal bowing (arrowhead); thickening of the free Rt. V. wall (arrow); and dilatation of the Rt atrium (*RA*). (b) Enhanced CT scan at a lower level than a shows dilatation of the Rt V. and IVC as well as a small pericardial effusion (*).



Fig 12. Severe pul. hypertension and Rt heart disease in a 75 y old patient.

CT scan shows opacification of the IVC and supra-hepatic veins because of retrograde flow of contrast material, which is often seen in patients with elevated Rt atrial and Rt ventricular pressures.

2-PA. aneurysm & pseudoaneurysm

-Congenital and acquired (both are rare).

-Causes → PDA, mycotic aneurysm, trauma, penetrating injury, Behçet disease, Takayasu disease, Marfan syndrome & pulmonary hypertension.

X-Ray → hilar enlargement.

CT → saccular / fusiform dilatation, homogeneous enhancement.

Behçet disease

-It is a chronic inflammatory disorder of unknown origin characterized by recurrent oral and genital ulcers, ocular anomalies and additional clinical manifestations in multiple organ systems.

-It is now recognized to be a systemic vasculitis that involve arteries & veins of any size → aneurysm / occlusion.

-Aneurysms are single / multiple.

X-Ray → enlarged hilum / round perihilar opacity.

CT → aneurysm +/- mural thrombus.

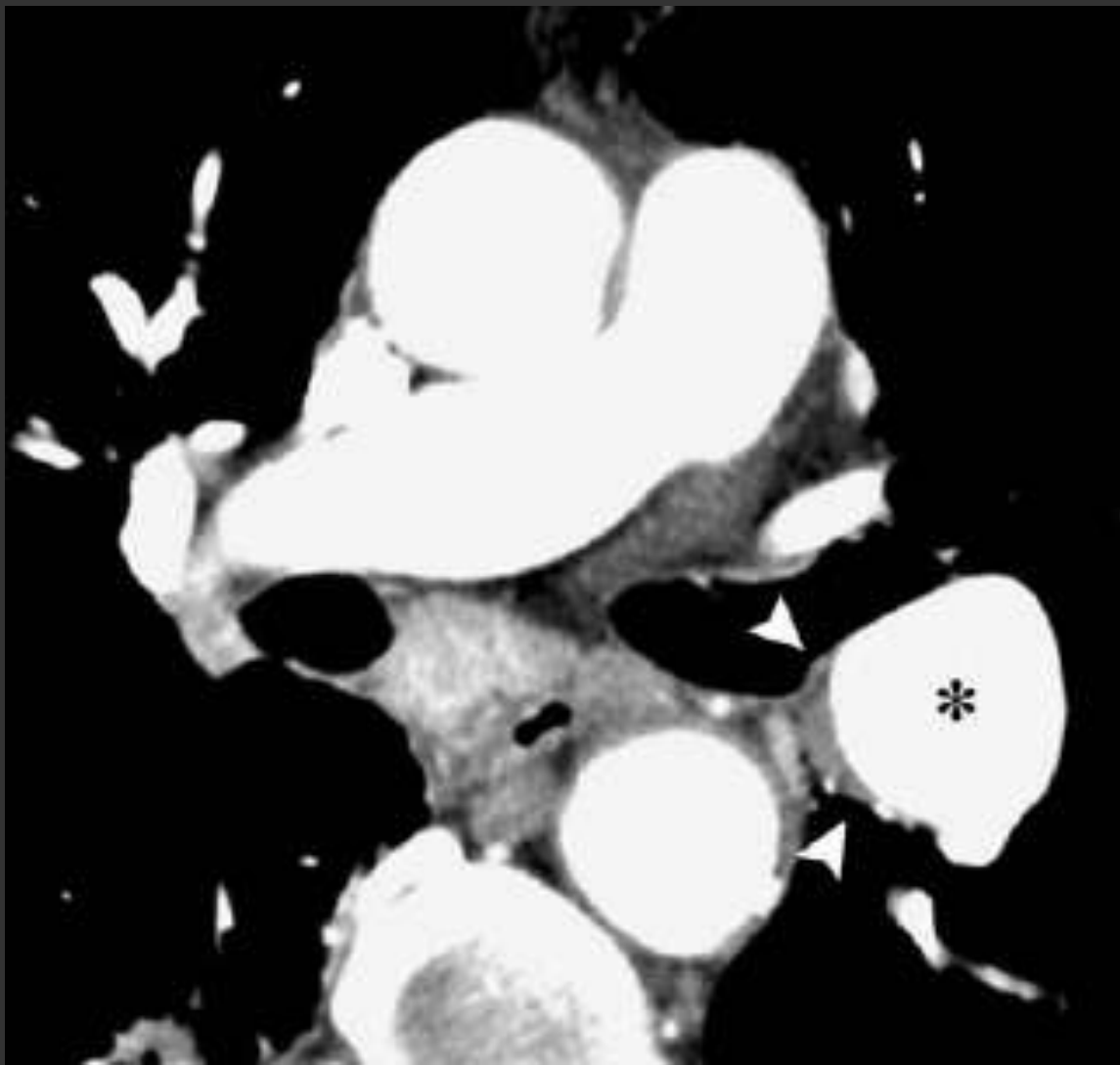


Fig 13. P.A. aneurysm in a 50 y old man with Behçet disease and hemoptysis.

Enhanced CT scan shows aneurysmal dilatation of a Lt interlobar P.A. (*) with small mural thrombi (arrowheads).

Mycotic (infected) aneurysm

- 2ry to endovascular seeding from septic pulmonary emboli / endocarditis.
- Single / multiple.
- Central / peripheral.



Fig 14. Septic emboli in a 20 y old woman with osteomyelitis and hemoptysis.

(a) CT scan at level of lower lobes shows an enhancing round lung mass with density similar to that of central arteries (not shown), findings that indicate an aneurysm in the Rt lower lobe artery (*). Peripheral triangular opacities in the Lt lung and a bilateral pleural effusion also seen. (b) CT scan (lung window) shows a cavitated peripheral nodule in Rt lung (arrowhead), enlargement of a Rt interlobular artery, and Rt sided pneumothorax (*) due to cavitory lesions. (c) MRA shows the Rt lower lobe P.A. aneurysm.

Rasmussen aneurysm

- Rare condition usually presented by hemoptysis.
- Erosion of P.A. wall by adjacent T.B. cavity.
- Usually peripheral.

X-Ray → intracavitary protrusion.

/→ replacement of a cavity by a nodule.

/→ a rapidly growing mass

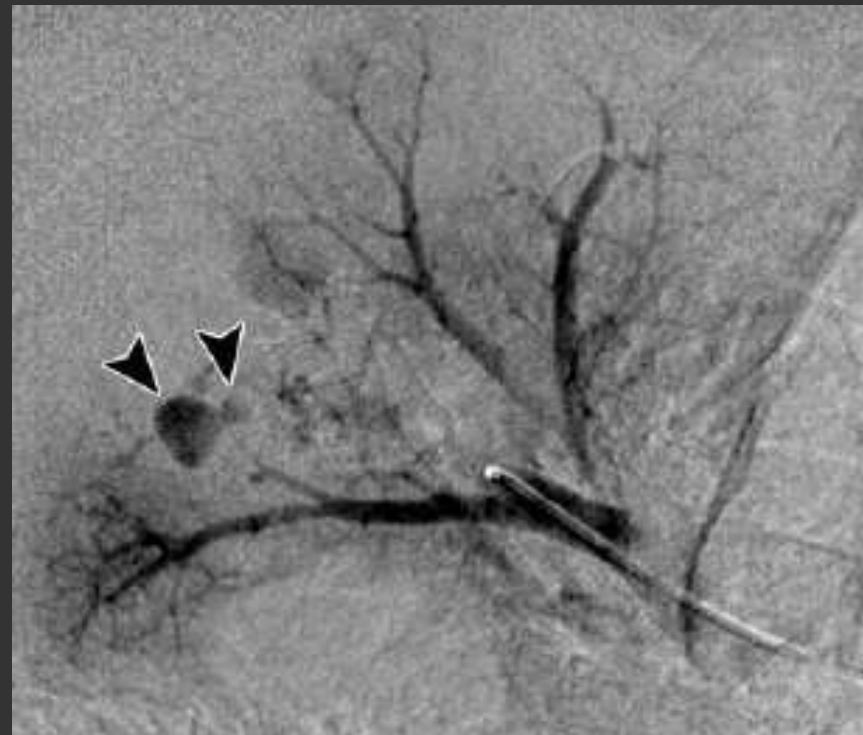


Fig 15. Rasmussen aneurysm in a 42 y old man with active postprimary T.B. and massive hemoptysis.

(a) Enhanced CT scan at level of upper lobes shows in an area of cavitation, a small rounded bilobed enhancing lesion (arrows) that arises from a branch of the P.A. (arrowhead). **(b)** Conventional angiogram shows contrast filling two aneurysms (arrowheads) in a segmental branch of the Rt upper lobe P.A.

3-Intravascular metastasis

-May involve main or segmental subsegmental arteries or arterioles.

-Symptoms vary and include progressive dyspnea, symptoms of subacute pulmonary hypertension or a more acute manifestation that mimics pulmonary thromboembolism.

CT → large emboli in main, lobar, segmental As → FDs.

→ small emboli in subsegmental As → multifocal dilatation /
beading of vessels.



Fig 16. Metastatic intravascular emboli in a 55 y old man with renal cell carcinoma.

CT scan (lung window) shows vascular dilatation and beading of subsegmental arteries (arrowheads), findings highly suggestive of metastatic intravascular emboli.

P.A. Narrowing

(↓ arterial diameter) (P.A. encasement & obstruction)

Include

1-Bronchogenic carcinoma.

2-Mediastinal fibrosis.

3-Takayasu arteritis.

1-Bronchogenic carcinoma

May encase MPA / proximal Rt or Lt P.A.

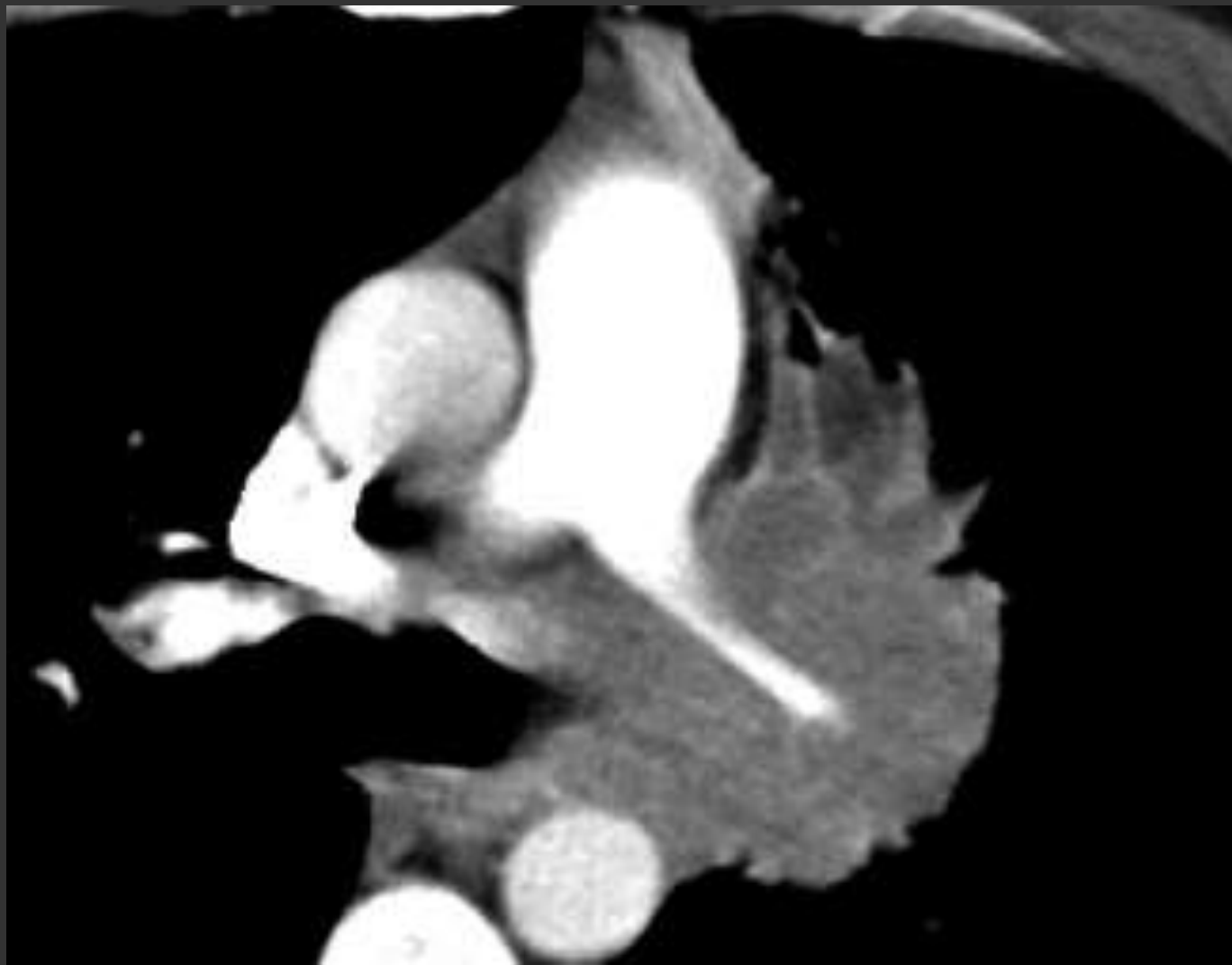


Fig 17. Unresectable bronchial carcinoma in a 30 y old man.

Enhanced CT scan shows extensive mediastinal tumor infiltration with obliteration of fat planes and encasement of the Lt P.A.

2-Mediastinal fibrosis

-Uncommon benign ch. inflammatory condition characterized by progressive proliferation of dense fibrous tissue in the mediastinum.

-Encase SVC, central airways, +/- P.A.

-**Focal type** → focal calcified mass in paratracheal / subcarinal / hilar regions (histoplasmosis / T.B).

-**Diffuse type** → diffuse infiltrative non calcified mass.

→ occurs with autoimmune diseases, retrop.

fibrosis, sclerosing cholangitis, drugs, idiopathic.

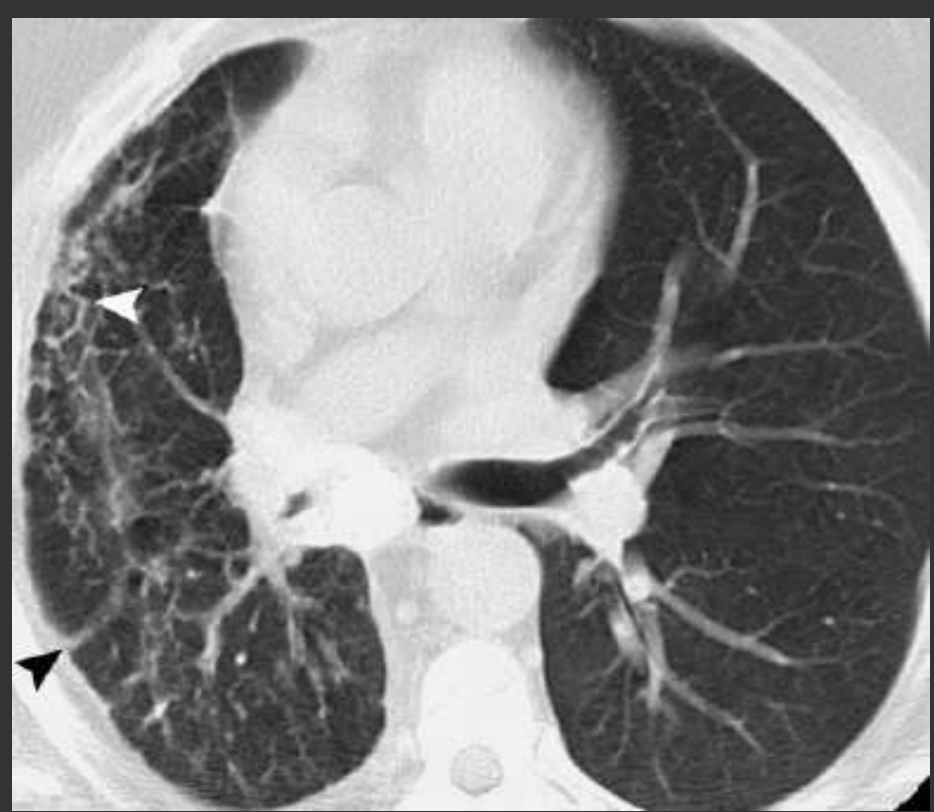


Fig 18. Focal mediastinal fibrosis 2ry to T.B. in a 62 y old man.

(a) Unenhanced CT scan at level of Rt hilum shows a highly calcified mass that encases Rt P.A. (curved arrow) and involves Lt lower lobe vein (straight arrow). (b) CT scan (lung window) shows multiple linear opacities perpendicular to the pleural surface (black arrowhead), enlarged septa (white arrowhead) due to systemic supply by collateral vessels (2ry to involvement of the Rt P.A.), and enlarged septal veins (2ry to involvement of the Lt lower lobe vein). Note the striking contrast in appearance between the Rt & Lt lungs.

3-Takayasu Arteritis

-Idiopathic arteritis mainly affect elastic As (aorta, its major branches, P.A.).

-Common in young Asian women.

CT → early → thick wall, enhancement, mural calcification.

→ later → luminal stenosis / occlusion of segmental &
subsegmental As.



Fig 19. Late-stage Takayasu arteritis with Rt P.A. involvement in a 63 y old woman.

(a) Unenhanced CT scan shows marked stenosis of the Rt P.A. (arrowheads), Lt sided pulmonary hypertension and wall calcification of the Lt P.A. and the ascending and descending aorta.



Fig 19. Late-stage Takayasu arteritis with Rt P.A. involvement in a 63 y old woman.

(b) Enhanced CT scan at the level of the supra-aortic trunks shows soft tissue that surrounds the brachiocephalic trunk (straight arrows), occlusion of Lt carotid artery (curved arrow), poor visibility of vessels in the Rt lung because of Rt P.A. involvement, and collateral vessel development from intercostal arteries (arrowheads). **(c)** Enhanced CT scan shows Rt P.A. occlusion (straight arrow), enlarged bronchial arteries (curved arrow) in the Rt hilum, and enlarged Rt internal mammary artery (arrowhead).

Other Acquired P.A. Lesions

P.A. Thrombosis

-Thromboembolism

-Nonthrombotic causes → fat, air, nitrogen, tumors, foreign
bodies, 1ry P.A. sarcoma.

1-Pulmonary thromboembolism

a-Acute (CTA)

1-Enlarged A. + complete FD + no enhancement.

2-Partial FD + marginal luminal contrast filling "railway track" sign.

3-Peripheral FD that forms acute angle with arterial wall.

-Parenchyma → peripheral wedge shaped infarction (typically in lower lobes).

-Cardiac → Rt. V. enlargement.



Fig 20. Acute occlusive pulmonary thromboembolism in a 58 y old woman.

Enhanced CT scan shows enlargement of an occluded artery (arrowheads) in the Lt upper lobe compared with the diameters of adjacent patent vessels (arrow), as well as filling defects in the Rt upper lobe artery.

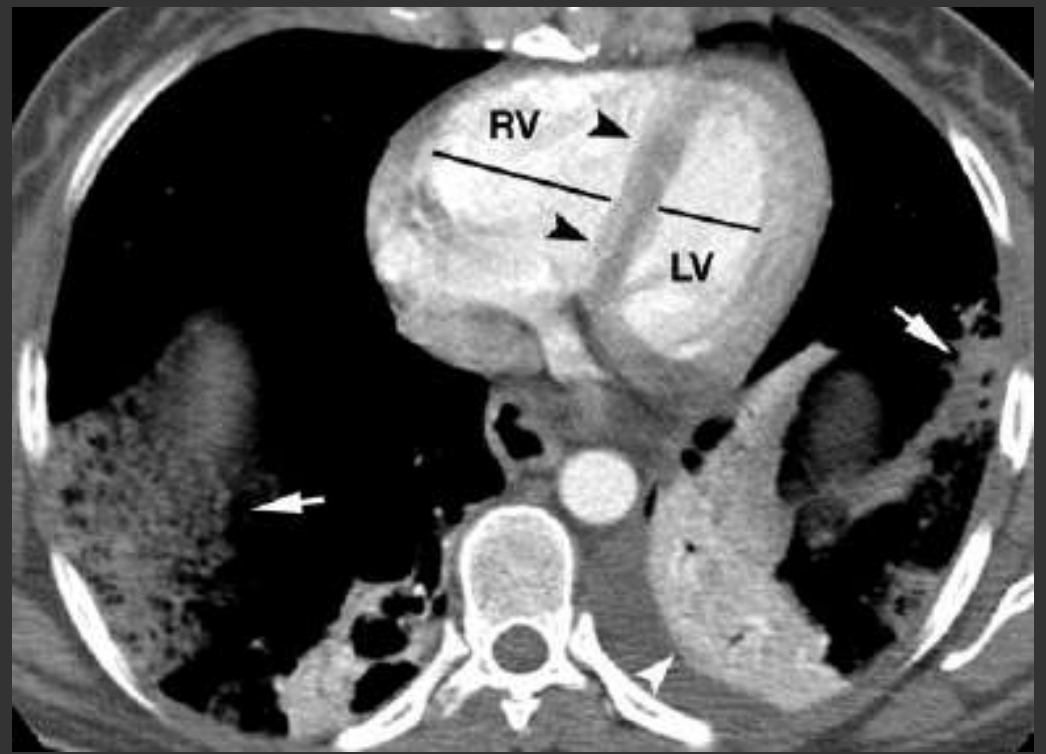
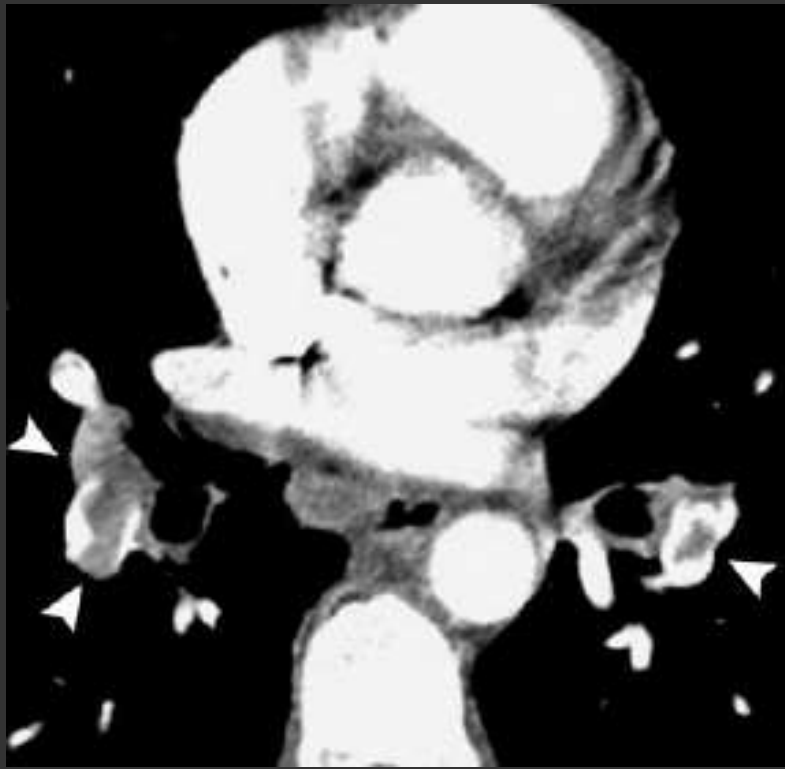


Fig 21. Acute pul. thromboembolism in a 32 y old woman with severe dyspnea.

(a) Enhanced CT scan shows significant bilateral partial FDs in peripheral segments of both interlobar arteries (arrowheads). In the Rt interlobar artery, the FD results in the "railway track" sign. In the Lt interlobar artery, the partial FD and surrounding area of contrast enhancement form acute angles with the arterial wall. (b) Enhanced CT scan at level of lower lobes shows peripheral triangular nonenhanced densities (arrows), suggestive of infarcts or haze, and some peripheral areas with enhancement (white arrowhead) suggestive of atelectasis. The short axis (black line) of the Rt. V. (*RV*) is wider than that of the Lt V. (*LV*), and mild displacement of the interventricular septa (black arrowheads) is visible. These abnormalities suggest Rt V. strain.

b-Chronic (CTA)

1-Peripheral flattened FD that forms obtuse angle with arterial wall (+/- calcification).

2-Retracted thrombus (complete FD at the level of stenosed A. with abrupt narrowing of the vessel).

3-Recanalization of thrombus (contrast flowing through As with thickened walls).

-Parenchyma → mosaic perfusion pattern.

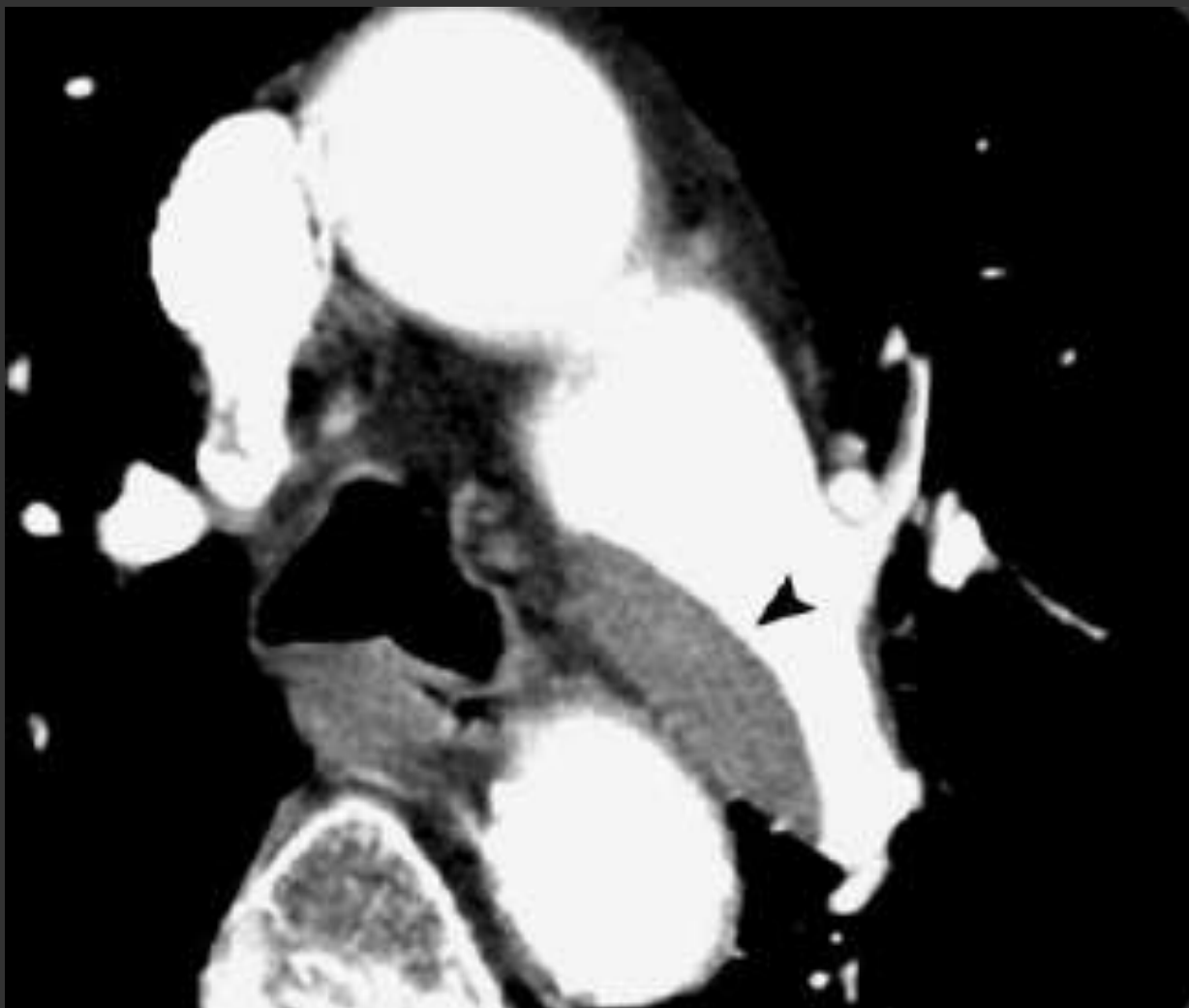


Fig 22. Ch. pulmonary embolism in a 62 y old man with dyspnea (same patient as in fig 9).

Enhanced CT scan shows a large eccentric pulmonary embolus (arrowhead) in the Lt main P.A.

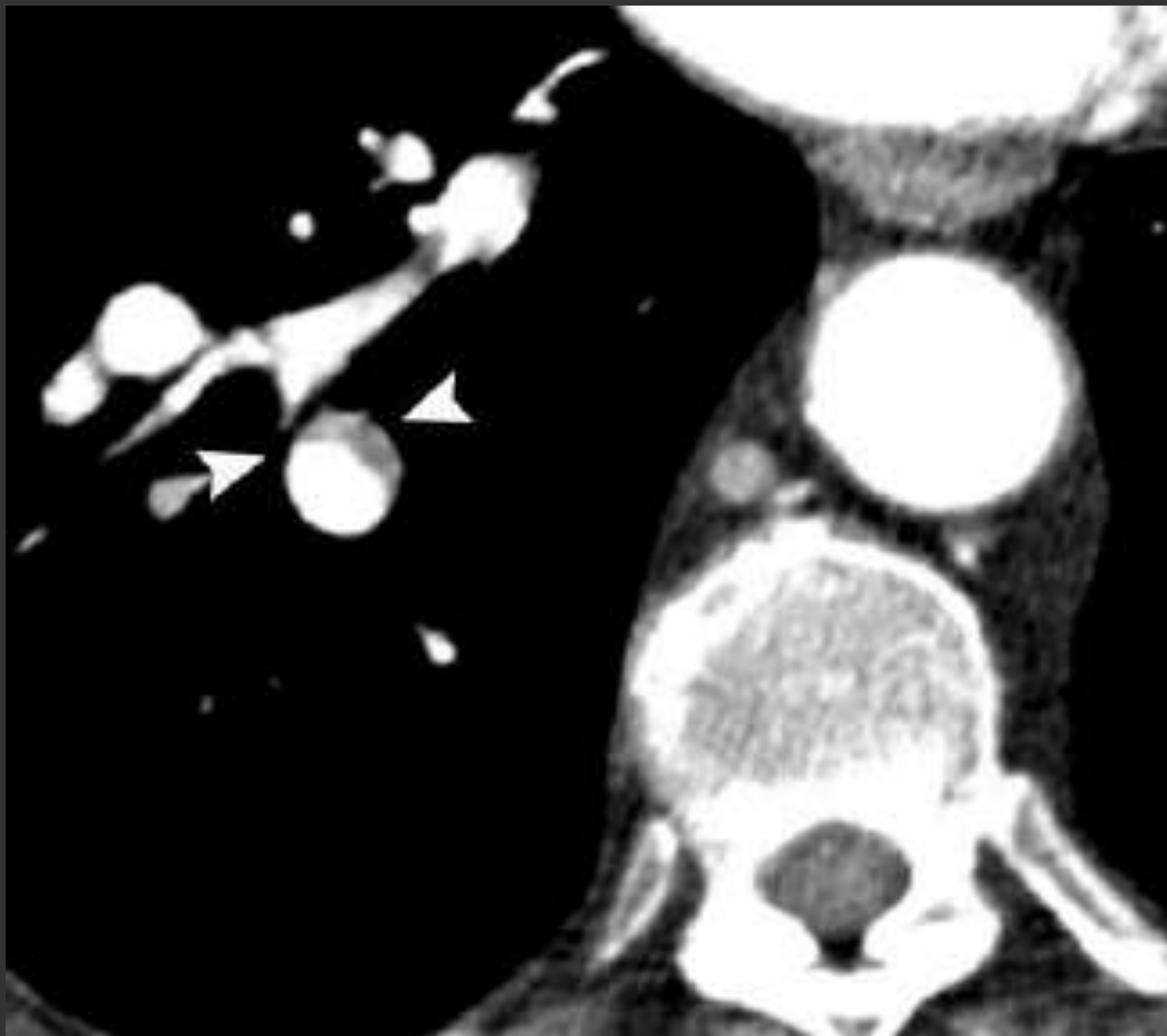


Fig 23. Ch. pulmonary embolism in a 72 y old woman.

Enhanced CT scan shows an eccentrically located small thrombus that forms obtuse angles with the vessel wall (arrowheads) in the Rt lower lobe.



Fig 24. Ch. pulmonary embolism in a 78 y old man.

(a) Unenhanced CT scan shows calcification at the level of Rt P.A. (white arrowhead) and a hypodense adjacent area (black arrowheads). Unlike acute pulmonary embolism, ch. pulmonary embolism may be indicated by hypodense clots at unenhanced CT. (b) Enhanced CT scan shows a peripheral clot in the corresponding hypodense area (arrowhead).

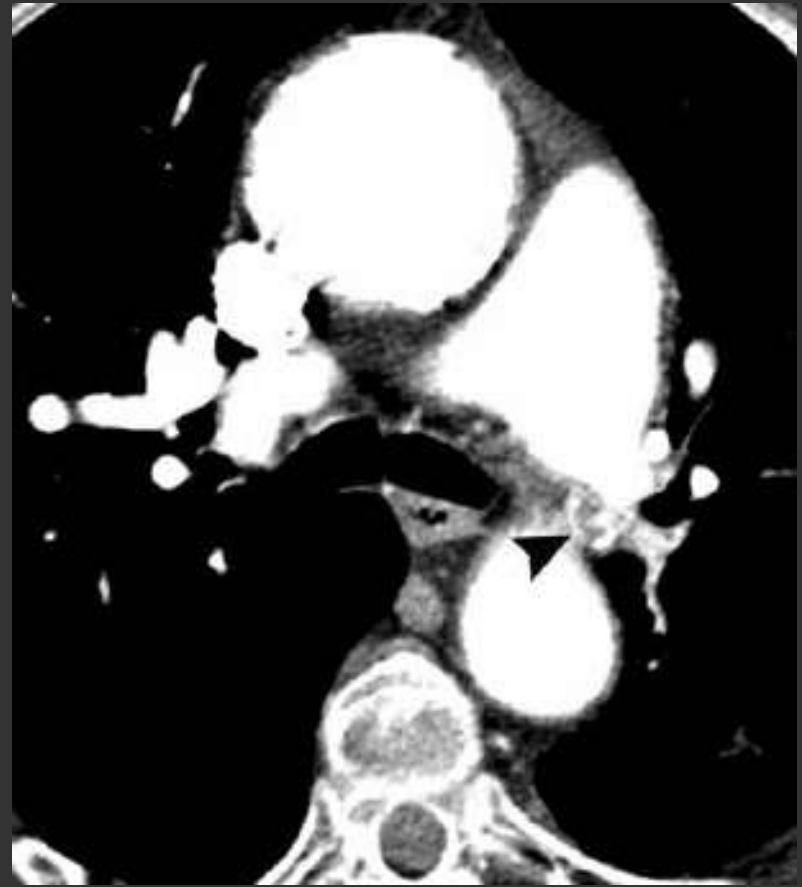
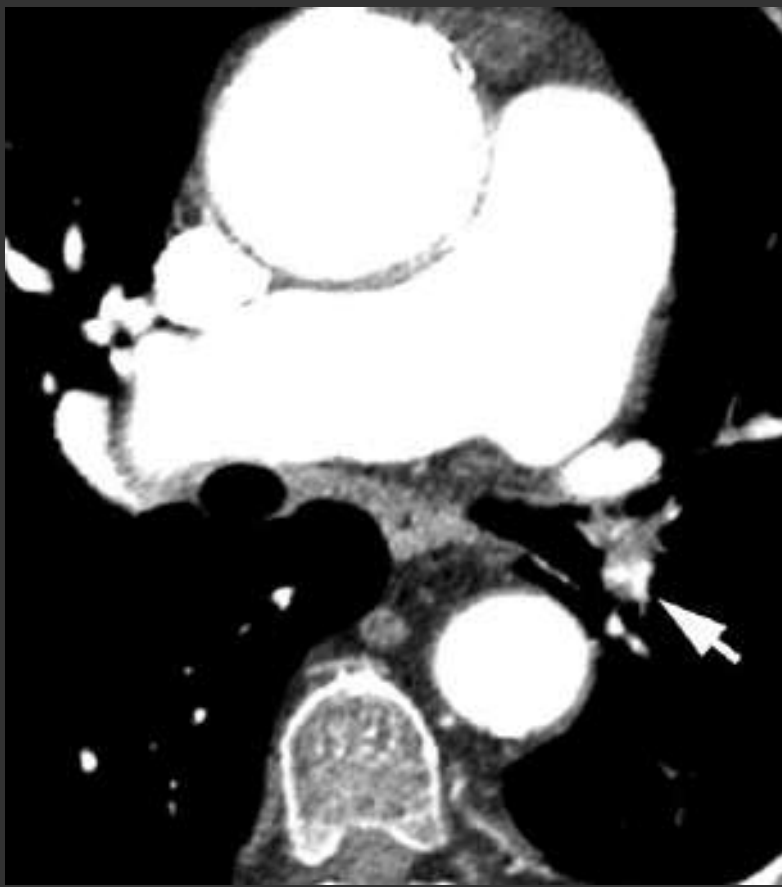


Fig 25. Ch. P.A. obstruction in a 68 y old woman with dyspnea.

(a) Enhanced CT scan at level of Lt hilum shows retracted embolic material (arrow); marked reduction in the diameter of Lt lower lobe P.A., and contrast material in the central lumen (finding suggestive of recanalization of the artery). **(b)** Enhanced CT scan at a level slightly higher than a shows enlargement of the bronchial arteries (arrowhead) because of a bronchial system-to-pulmonary system shunt.



Fig 25. Ch. P.A. obstruction in a 68 y old woman with dyspnea.

(c) Unenhanced CT scan (lung window) at the level of lower lobes shows marked stenosis of Lt lower lobe arteries and dilatation of the accompanying bronchi (arrow); multiple linear opacities (arrowheads) adjacent to the pleural surface in Lt lower lobe, suggestive of transpleural systemic vessels; marked dilatation of vessels in Rt lung; and a thrombus (*) in Rt lower lobe artery.

P.A. sarcoma

- Undiff. sarcoma or leiomyosarcoma.
- The main or proximal P.As. are most frequently involved.
- Intraluminal FD (mimics thromboembolus) but it fills, spans and expands the entire lumen of MPA / proximal PA (this finding is unusual in pulmonary thromboembolism).
- Extension into mediastinum / lung.
- Delayed enhancement.

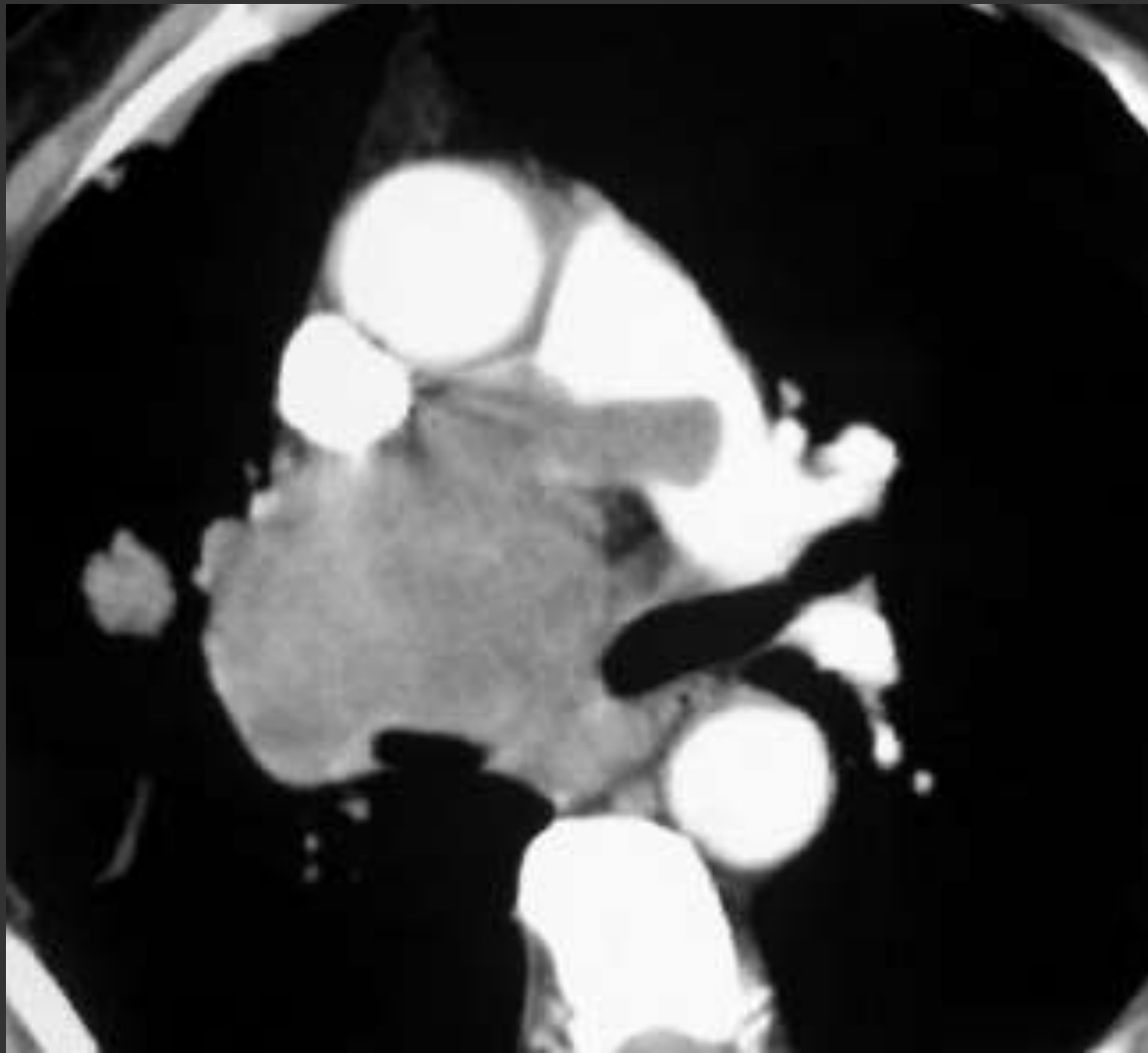


Fig 26. P.A. sarcoma in a 70 y old man with dyspnea.

Enhanced CT scan shows FDs in the main, Lt & Rt P.As. and the Rt interlobar P.A. The arterial lumina are expanded and there is extravascular invasion.

Conclusion

-Most P.A. anomalies can be detected with chest radiography.

-Definitive diagnosis can be reached with CT.

-Radiologists should be aware with different entities that may affect P.A.

-It is important to recognize the CT signs of these conditions to help correct diagnosis.

Thank You