PICTORIAL REVIEW OF PULMONARY VASCULATURE : FROM ARTERIES TO VEIN

Presenter : Dr Abdul Hadi

Supervisor : Dr Nasibah Mohamad

Introduction

- Pathology of the pulmonary vasculature involves an impressive array of both congenital and acquired conditions.
- Some of benign but some pulmonary vasculature is often incompatible with life.
- Although many of these conditions are rare, efficient and accurate diagnosis are essential as many of these disorders are life-threatening.
- This review present a guide to disorders of the pulmonary vasculature, covering the congenital and acquired conditions of the pulmonary arteries, pulmonary veins and bronchial arteries.
- The goal is to provide an integrated overview of the features of all major disorders of the pulmonary vasculature
- Assist the radiologist in identifying pathology
- Make differential diagnosis tailored to the presenting patient.

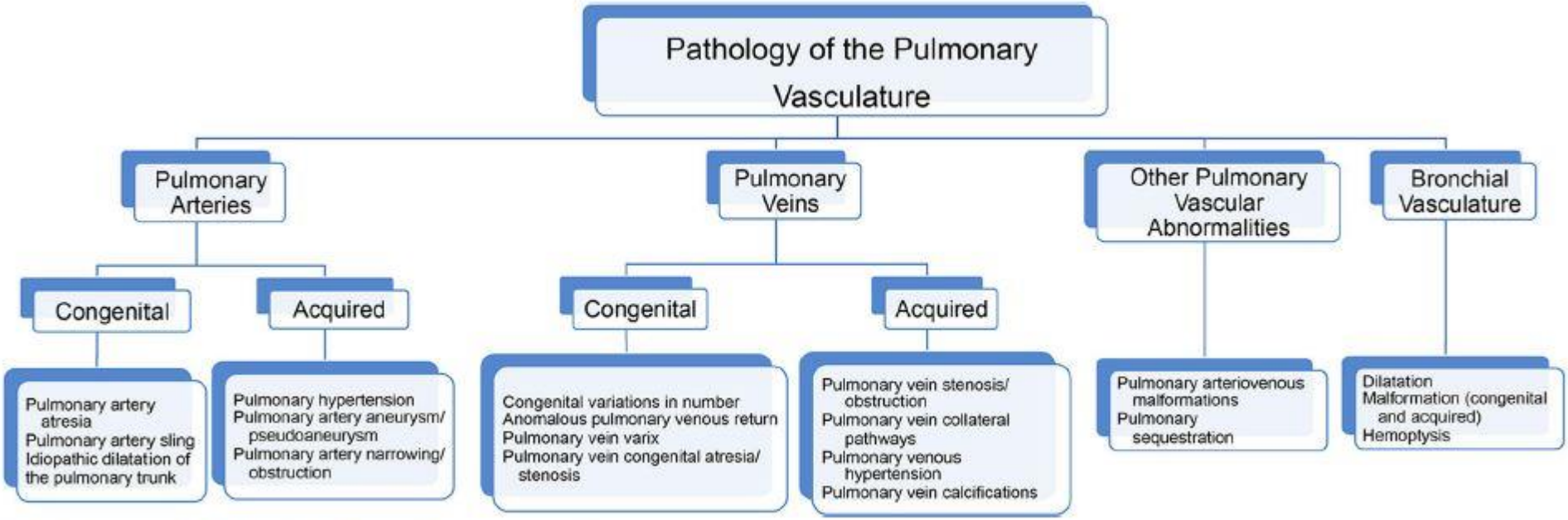


Fig. 1 Our conceptual organisation of pulmonary vascular pathology

- Chest radiograph have limited role is assesment potential for these conditions.
- CT is the gold standard for evaluating most vascular pathology.
  - Ability clearly elucidate anatomical detail
  - Good in detailed look at complex pulmonary vascular anatomy.
- In infant or children, ultrasound is first line modalities with subsequent follow up by MRI to reduce unnecessary radiation.

Pulmonary Artery Abnormality

- CONGENITAL DISORDER OF PULMONARY ARTERY

1. Pulmonary Artery Atresia

- Failed development of pulmonary artery → pulmonary artery atresia
- AKA proximal interruption of the pulmonary artery
- Distal part of the pulmonary artery are often intact and perfused due to collateral flow from aortopulmonary collateral vessels
- Rt pulmonary artery atresia : no associated congenital disease
- Lt pulmonary artery atresia : associated congenital disease eg. TOF
- Usually symptomatic : dyspnea, recurrent infection, hemorrhage.

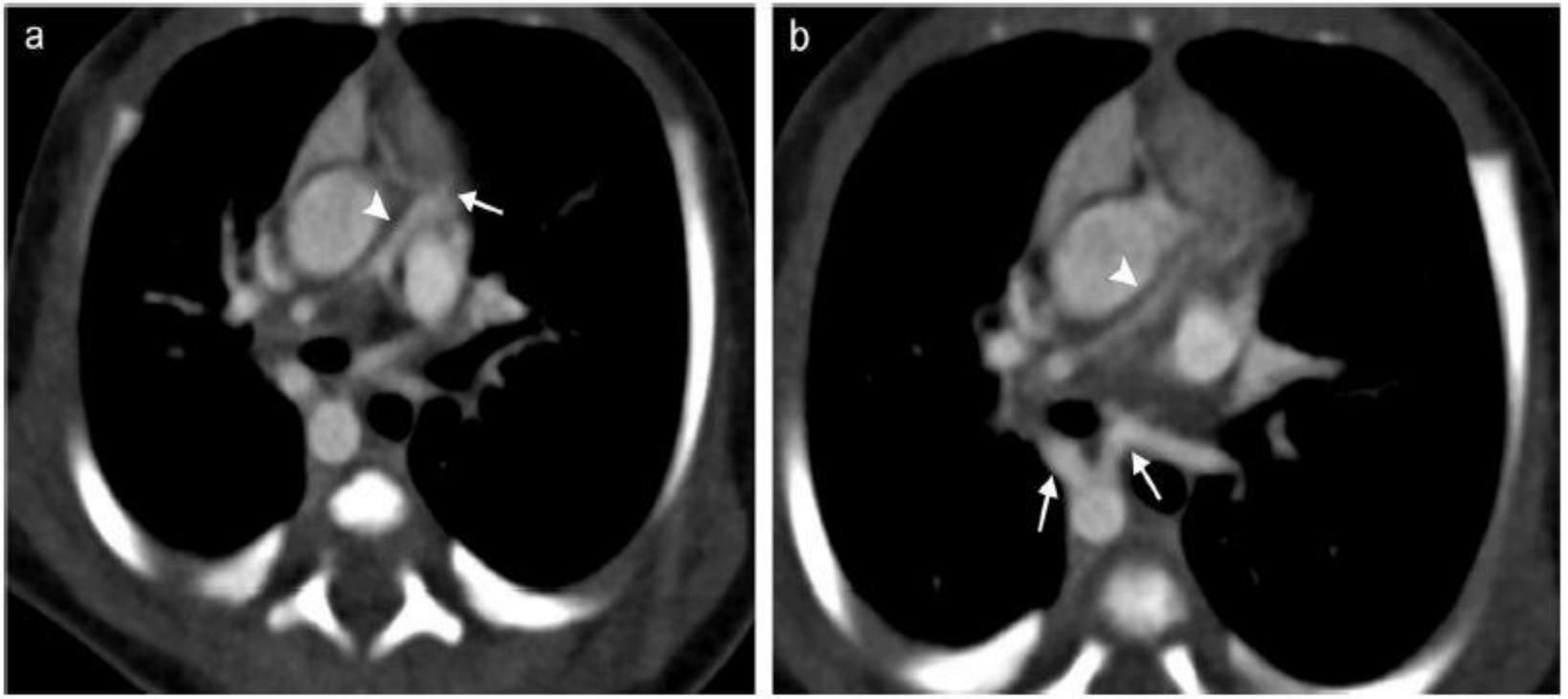


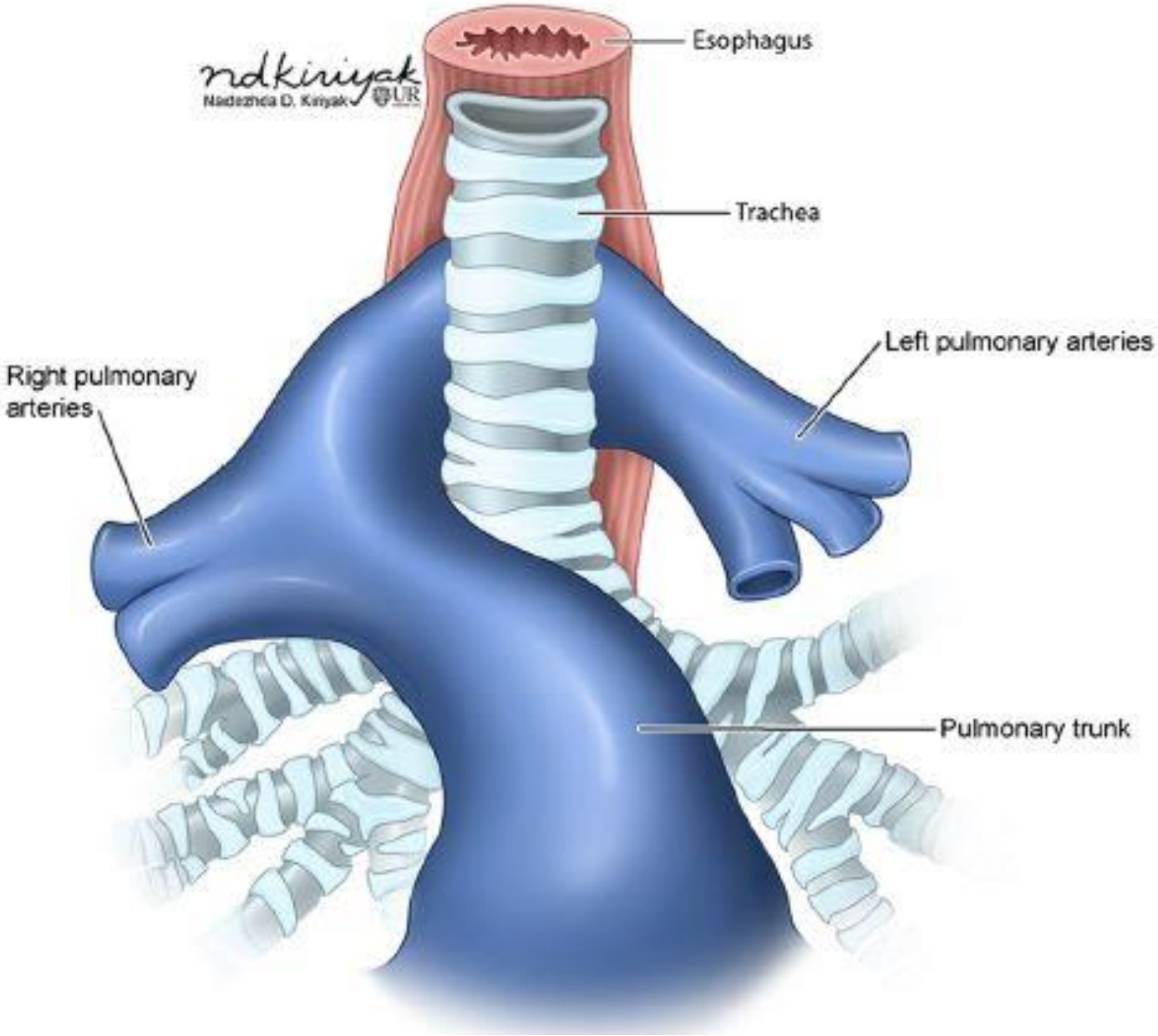
Fig. 2 Pulmonary artery atresia. a Axial CT image demonstrating abrupt termination of the main pulmonary artery (*white arrow*) in a patient with tetralogy of Fallot with a small hypoplastic distal pulmonary artery (*white arrowhead*). b Bronchial artery dilatation off the descending aorta occurs to feed the pulmonary vasculature (*white arrows*) with the hypoplastic distal pulmonary artery again demonstrated (*white arrowhead*)

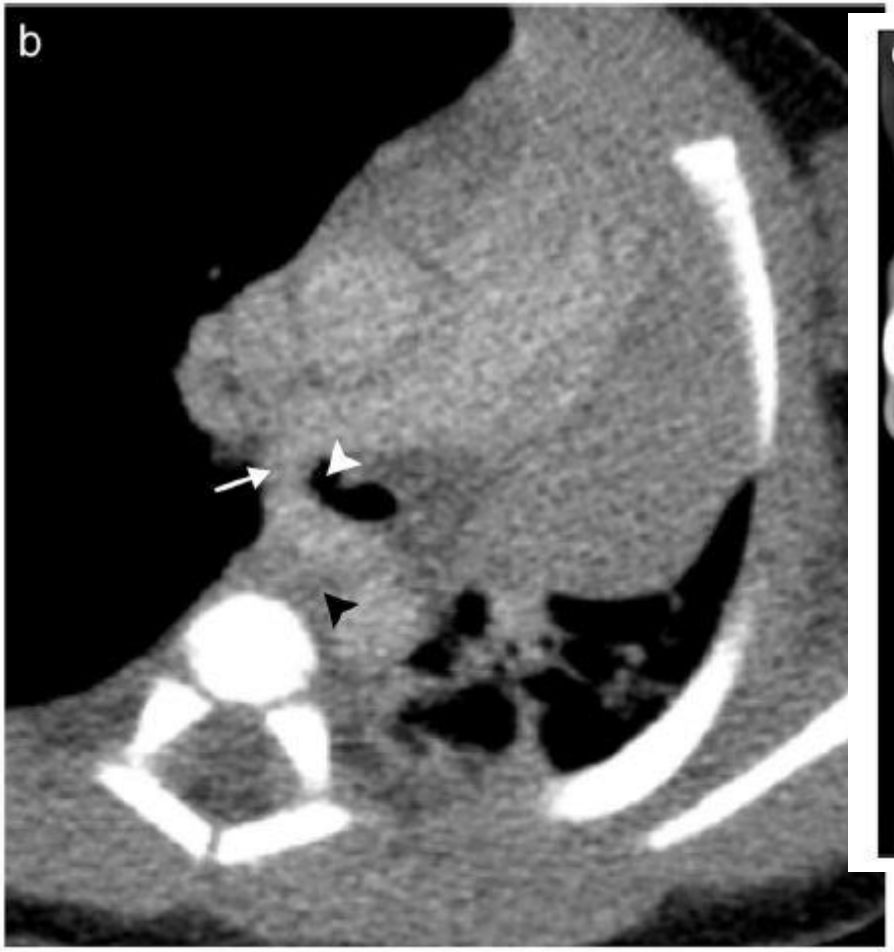
- Absence or hypoplasia of affected proximal artery
- Additional findings : hypoplasia of the affected lung, serration and thickening of the pleura and aortopulmonary collateral (APC) vessel formation

Pulmonary Artery Abnormality

2. Pulmonary Artery Sling (Aberrant left pulmonary artery)

- Developmental failure of the left sixth aortic arch
- Arising from the right pulmonary artery → courses between the trachea/right mainstem bronchus and oesophagus
- Can cause compression/focal stenosis of the airway → lung atelactasis
- Most of the patient asymptomatic due to only mild compression.
- Plain CXR : left-sided deviation of the trachea and mediastinum.
- Barium swallow : anterior indentation of the oesophagus.


ndkiriyak
Nadezhda D. Kiriyak
UR
Esophagus
Trachea
Left pulmonary arteries
Right pulmonary arteries
Pulmonary trunk



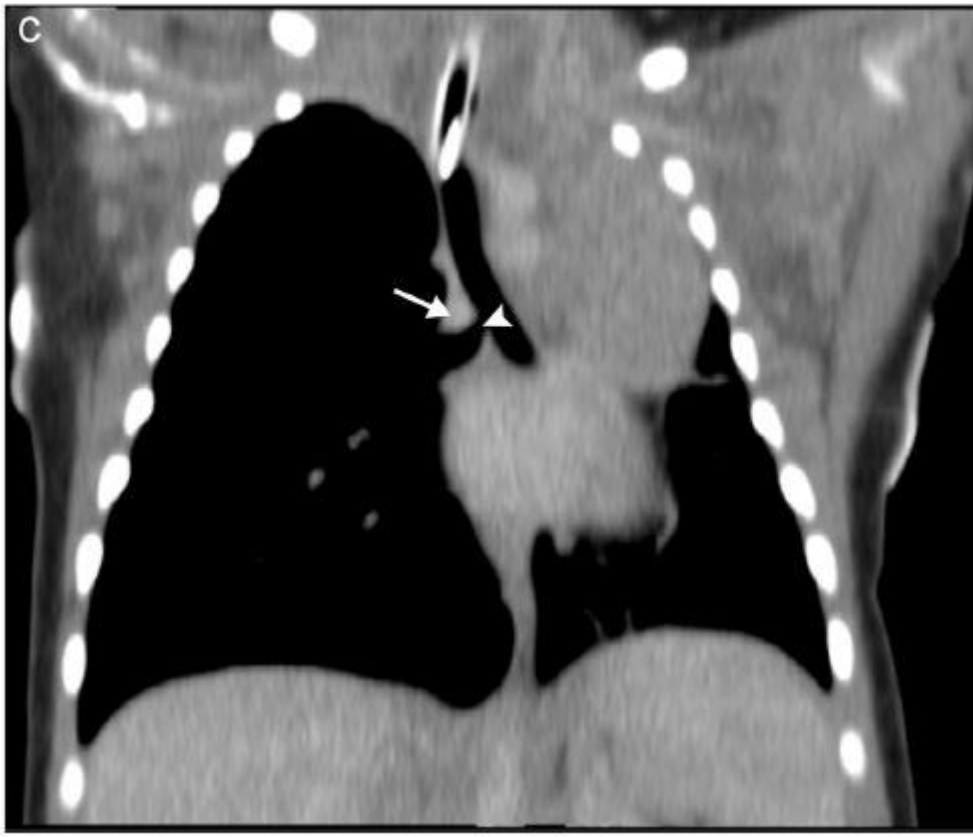


Fig. 3 Pulmonary artery sling. Schematic (**a**) and axial contrast-enhanced CT image (**b**) show the left pulmonary artery (*white arrow*) arising from the right pulmonary artery, passing between the oesophagus (*black arrowhead*) and right mainstem bronchus (*white arrowhead*). **c** Coronal maximal intensity projection CT image shows compression of the proximal right mainstem bronchus (*white arrowhead*) by the aberrant left pulmonary artery (*white arrow*)

Pulmonary Artery Abnormality

3. Idiopathic dilatation of the pulmonary trunk

- Diagnosis of exclusion.
- Congenital dilatation of pulmonary trunk with normal pressure in the absence of other cardiopulmonary disease.
- Asymptomatic patient although close follow up is recommended.
- Transverse axial diameter of the pulmonary trunk can be measured at the level of the bifurcation of the right pulmonary artery

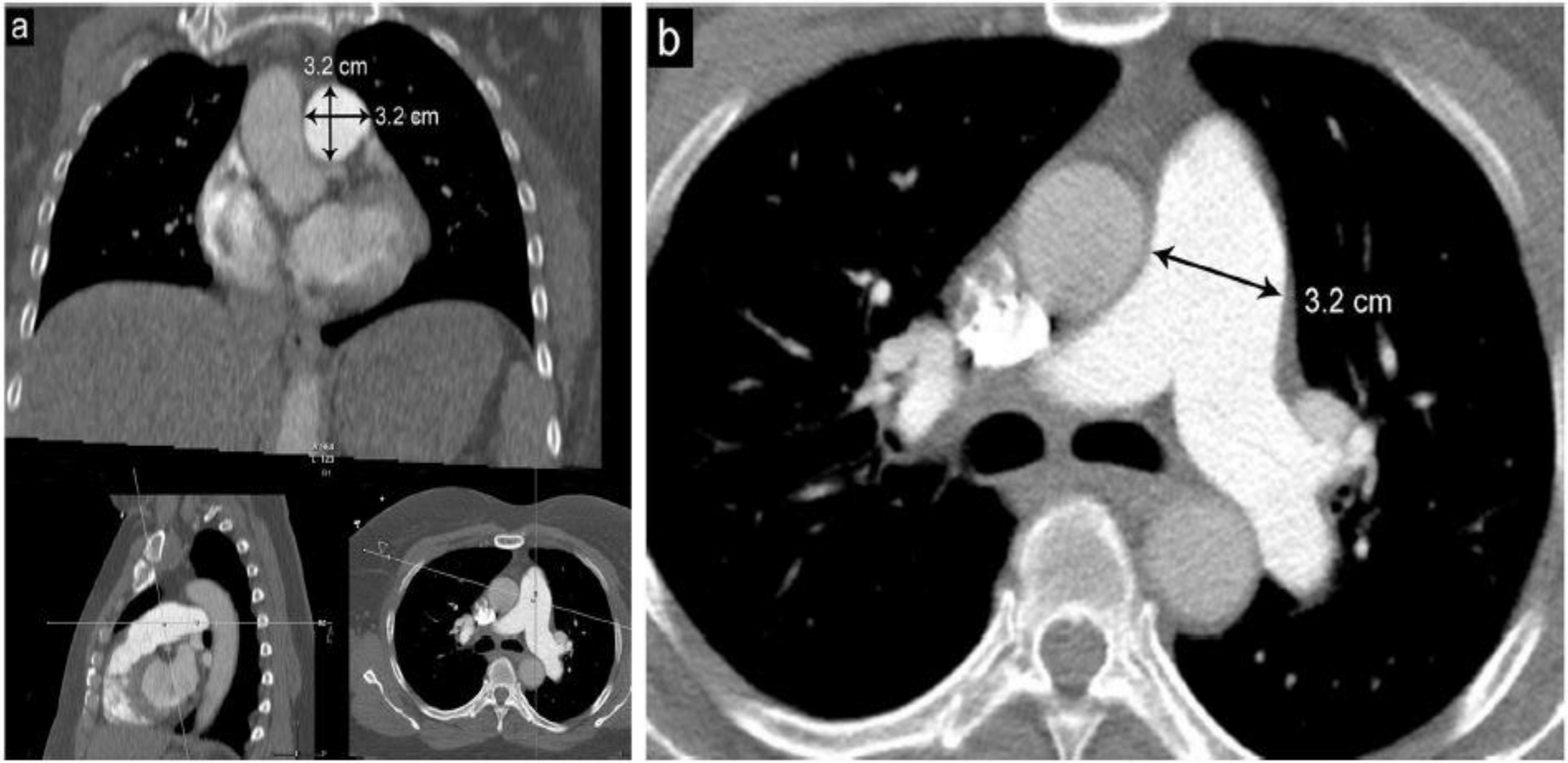


Fig. 4 Pulmonary artery measurement. **a** CT imaging demonstrating double oblique measurement technique through the main pulmonary artery. **b** Axial CT image demonstrating transverse measurement through the main pulmonary artery at the level of the right pulmonary artery bifurcation

Pulmonary Artery Abnormality

- ACQUIRED DISORDER OF PULMONARY ARTERY

1. Pulmonary hypertension

- PA pressure >25mmHg at rest(assesed by right heart artery catheterisation).
- Rarely idiopathic. Commonly secondary to other pathology :
  - Heart failure
  - Pulmonary parenchymal disease (including fibrosis and emphysema)
  - Chronic pulmonary emboli
- Imaging findings :
  - Enlarged pulmonary trunk (>29mm)
  - pulmonary artery diameter to ascending aorta diameter of > 1
  - segmental artery-to bronchus ratio > 1 in at least three of four lobes.

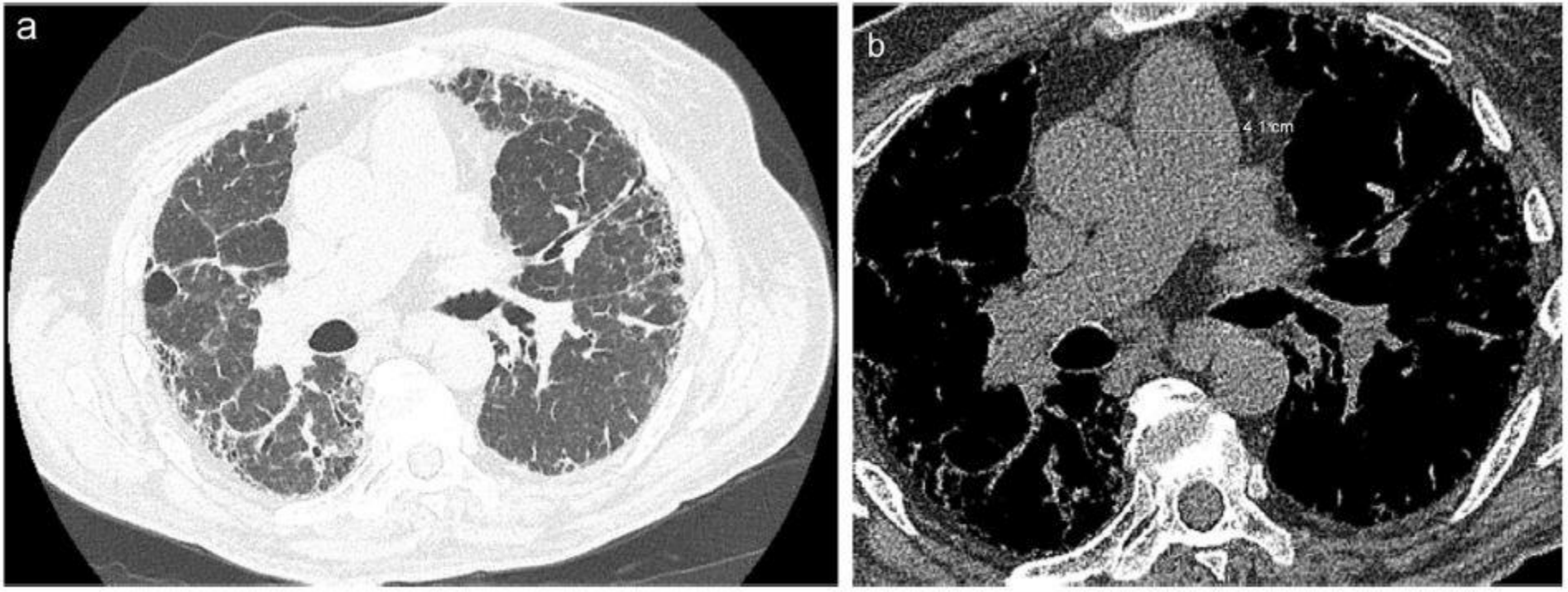


Fig. 5 Pulmonary hypertension in a patient with idiopathic pulmonary fibrosis. **a** Axial CT image (lung window) shows basal and peripheral predominant fibrotic disease with reticulation and honeycombing extending to the subpleural surface, as well as traction bronchiectasis, consistent with usual interstitial pneumonia pattern of fibrosis in a patient with idiopathic pulmonary fibrosis. **b** Axial CT image shows dilatation of the main pulmonary artery, measuring up to 4.1 cm in maximal diameter

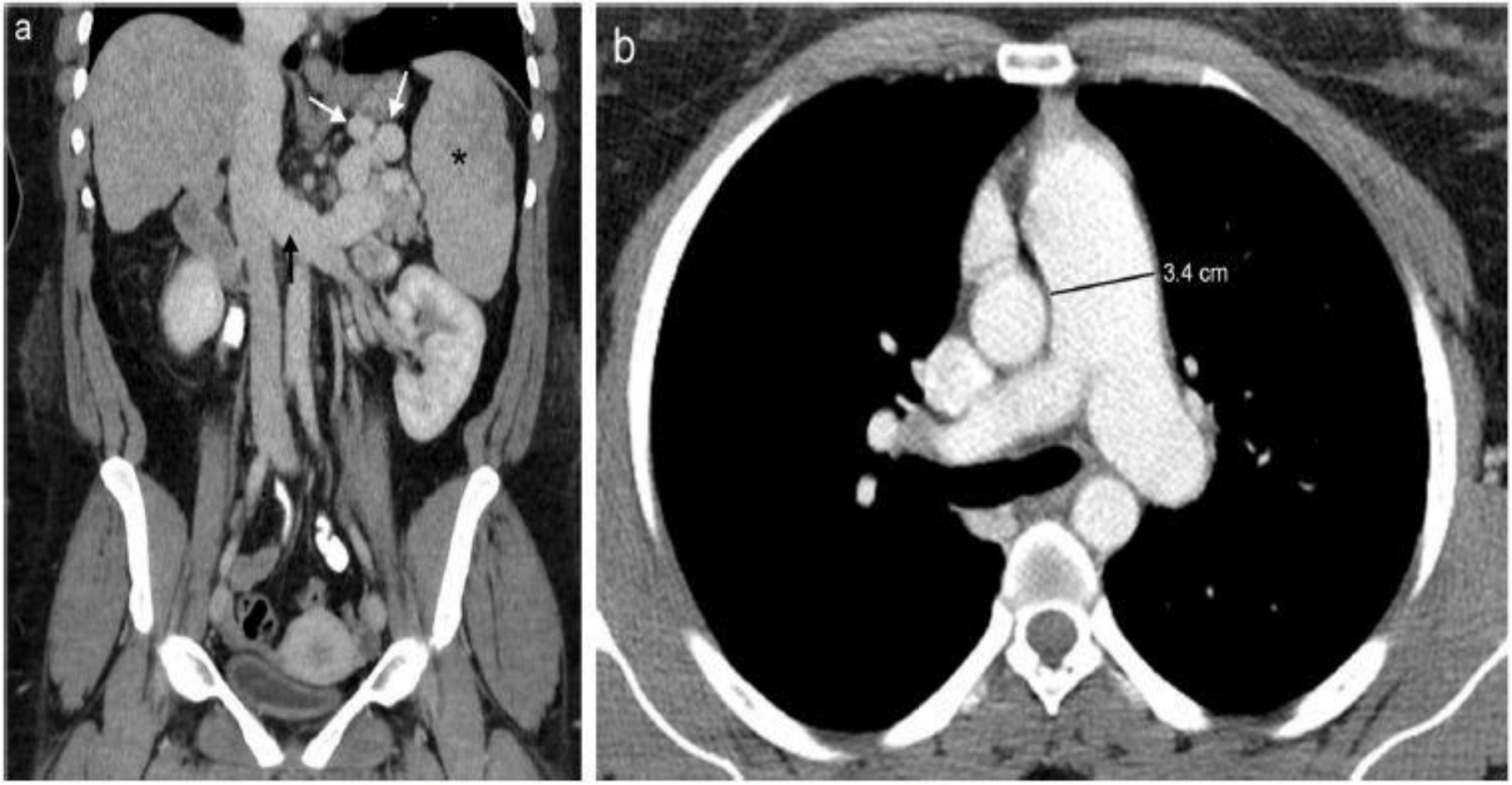


Fig. 6 Pulmonary hypertension in a patient with liver disease. **a** Coronal contrast-enhanced CT image shows sequela of portal hypertension secondary to cirrhotic liver morphology with splenomegaly (*), enlarged splenorenal collateral vessels (*white arrows*) and enlarged left renal vein (*black arrow*). **b** Axial contrast-enhanced CT image shows dilatation of the pulmonary arteries with the main pulmonary artery measuring up to 3.4 cm in maximal diameter

Pulmonary Artery Abnormality

2. Pulmonary artery aneurysm/pseudoaneurysm

- May arise secondary to
  - Pulmonary hypertension
  - Collagen/ vascular conditions (eg marfan syndrome, Takayasu arteritis and Behçet's disease)
  - Iatrogenic catheter misplacement
  - Trauma or infection
- May occur in isolation or may also associated with other congenital heart disease.
- Imaging may reveal :
  - Focal segment dilatation of pulmonary artery
  - Often saccular in appearance
  - Phase of contrast following pulmonary artery enhancement

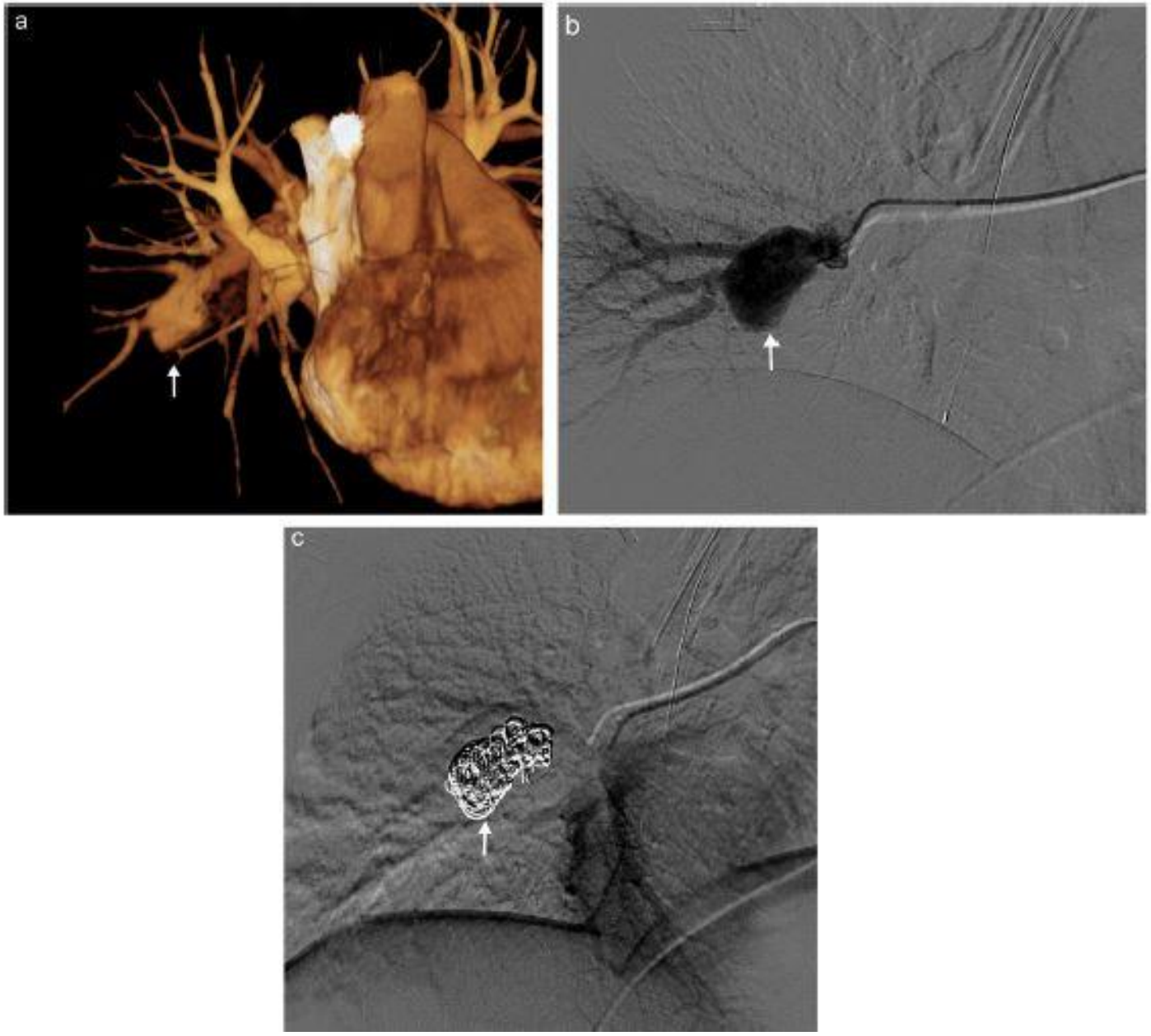


Fig. 7 Pulmonary artery aneurysm in a patient with Behçet's disease. **a** Three-dimensional reconstruction from a contrast-enhanced CT scan showing a right lower lobe pulmonary artery aneurysm in a patient with Behçet's disease (*white arrow*). Digital subtraction fluoroscopic images obtained before (**b**) and after (**c**) coil embolisation of the right lower lobe pulmonary aneurysm (*white arrow*)

Pulmonary Artery Abnormality

3. Pulmonary artery narrowing/obstruction

- Obstruction secondary to
  - Primary sarcoma of pulmonary artery
  - Pulmonary malinancy
  - Inflammatory condition eg. Mediastinal fibrosis, takayasu arteritis
  - Both acute and chronic pulmonary embolism

- Primary sarcoma of pulmonary artery a rare condition
  - Often mimicking the pulmonary embolism in CT findings
  - CT will shows enlarged artery with a filling defect that may encompass entire lumen
  - Other findings (to differentiate with thrombus):
    1. Entire involvement of proximal and main pulmonary arteries.
    2. Enlargement of the involved pulmonary arteries
    3. Extension into the surrounding tissue
    4. Small enhancing vessels within the tumor

Pulmonary Artery Abnormality

- Malignant primary neoplasm can cause external compression or directly invading the pulmonary artery
- Fibrosing mediastinitis : proliferation of fibrous tissue in mediastinum → obstruction and narrowing the pulmonary artery
- Takasayu arteritis : circumferential wall thickening and enhancement of pulmonary artery → obstruction and narrowing the pulmonary artery

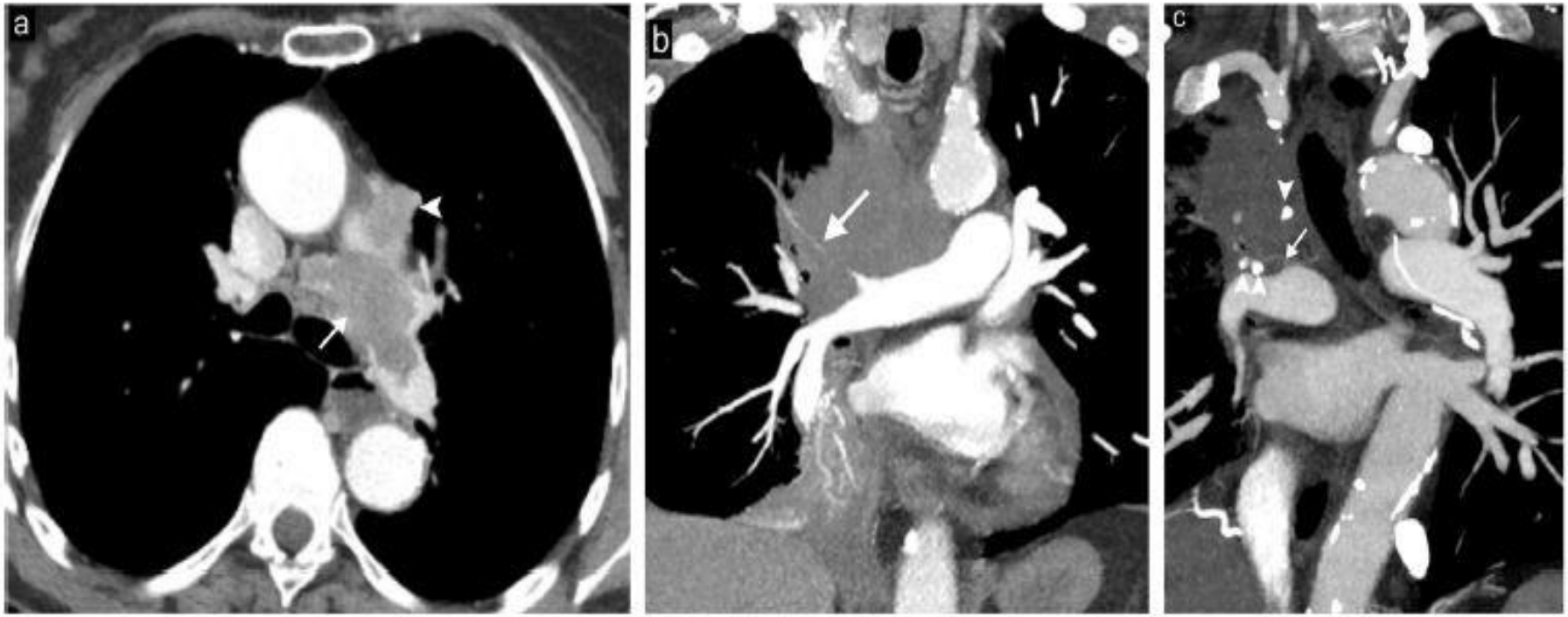


Fig. 8 Pulmonary artery narrowing/obstruction. **a** Axial contrast-enhanced CT image shows a large, irregularly shaped filling defect within the left pulmonary artery (*arrow*) with extension into the mediastinal fat (*arrowhead*) causing intrinsic narrowing/obstruction of the pulmonary artery in a patient with biopsy proven angiosarcoma. **b** Coronal CT image demonstrating severe narrowing of the right upper lobe pulmonary artery (*arrow*) secondary to extrinsic mass effect from a right upper lobe primary lung malignancy. **c** Coronal CT image demonstrating complete occlusion of the right upper lobe pulmonary artery (*arrow*) secondary to extrinsic compression from biopsy proven fibrosing mediastinitis. Note the coarse calcifications present within the region of fibrosis (*arrowheads*)

Pulmonary Artery Abnormality

- Pulmonary embolism : most common causes of pulmonary artery obstruction
  - Best evaluated with CTPA.
  - Demonstrate central focal filling defect , rimmed by contrast
  - Enlargement of affected branch may also seen

- Chronic pulmonary embolism
  - Embolus incorporated into the luminal wall → anticoagulant become ineffective → stenosis of pulmonary artery → pulmonary hypertension
  - CT findings : 1) Eccentric thrombus → partial or complete obstruction
    2) Partial calcification within thrombus
    3) Web band like within pulmonary artery
    4) May has calcifications
  - Indirect sign : Mosaic attenuation of pulmonary parenchyma due to perfusion defect or Aortopulmoary collateral formation

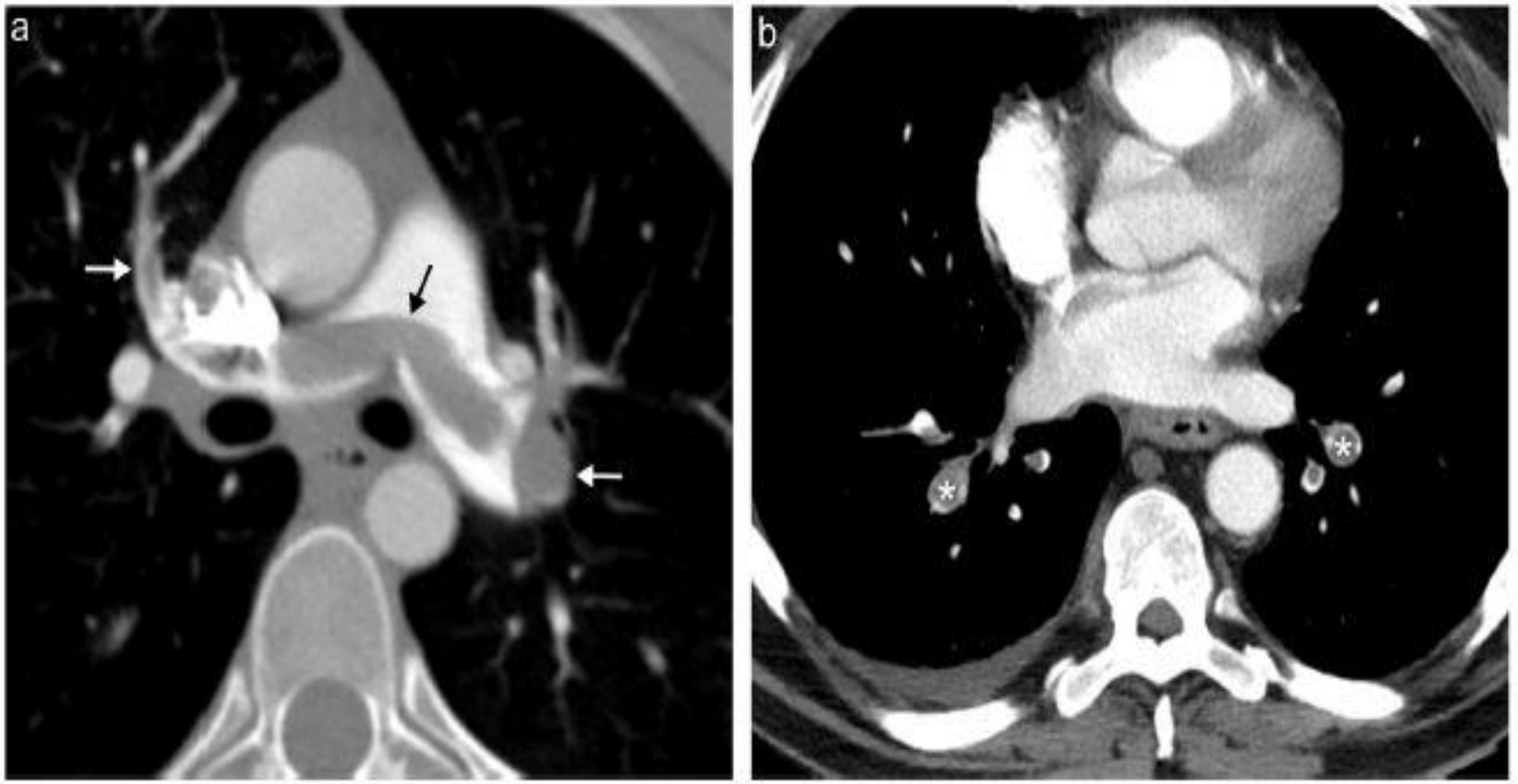


Fig. 9 Two patients with acute pulmonary emboli. **a** Axial contrast-enhanced CT image shows a large saddle embolus of the main pulmonary artery (*black arrow*) as well as lobar/segmental involvement of the right and left pulmonary arteries (*white arrows*). **b** Axial contrast-enhanced CT image of another patient with bilateral acute pulmonary emboli demonstrate the central focal filling defect within bilateral segmental pulmonary arteries which is rimmed by contrast (*)

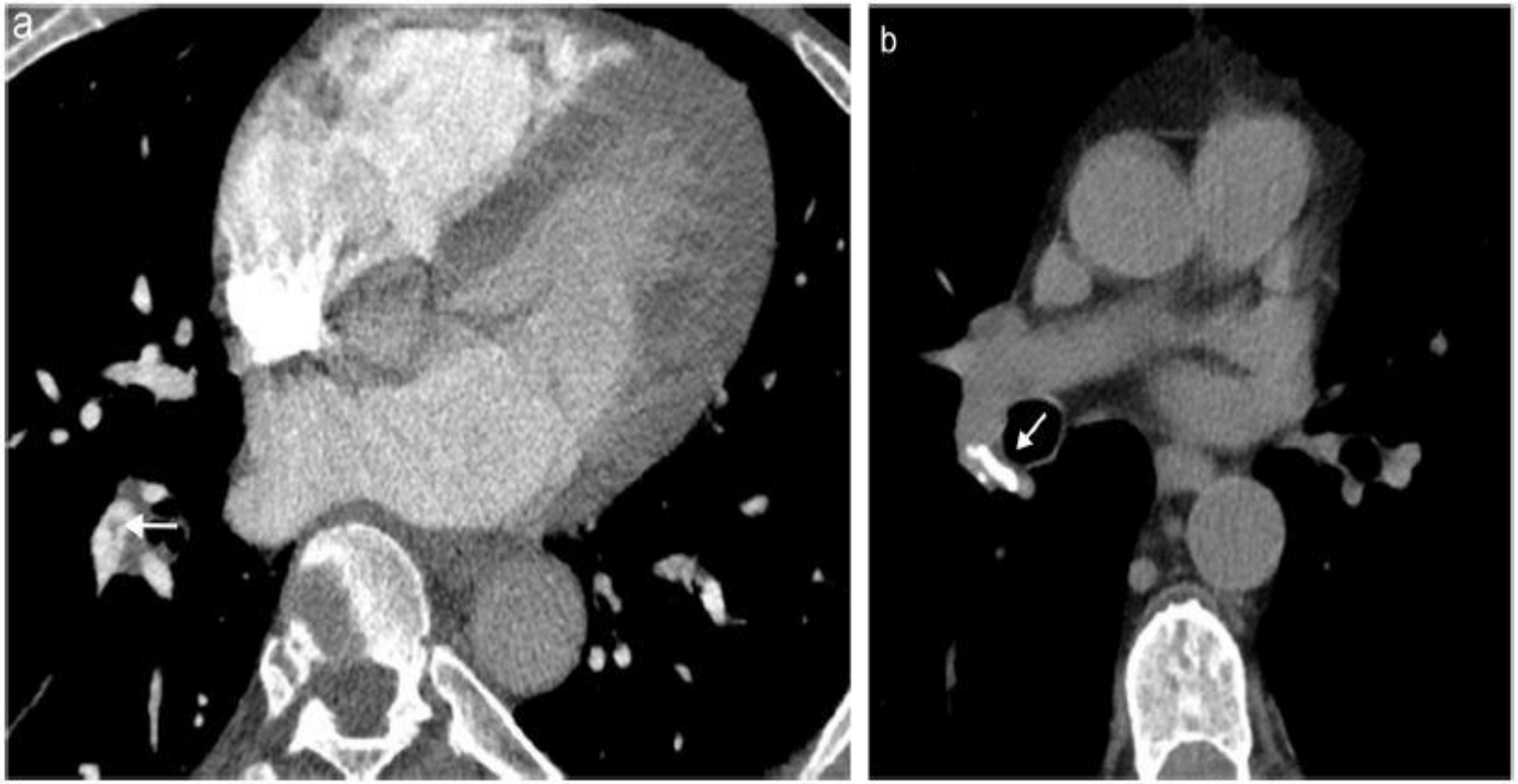


Fig. 10 Two patients with chronic pulmonary emboli. a Axial contrast-enhanced CT image shows a web-like filling defect within a segmental right lower lobe pulmonary artery consistent with chronic pulmonary embolus (*white arrow*). b Axial non-contrast CT image of another patient demonstrates linear calcification within the right interlobar pulmonary artery consistent with chronic pulmonary embolus (*white arrow*)

Pulmonary Vein Abnormality

- CONGENITAL DISORDER OF PULMONARY VEIN

1. Congenital variations in number

- Common : 4 pulmonary vein(PV) drain into Lt atrium (2 right and 2 left).
- Variant :
  - Common left pulmonary vein : Superior & Inferior PV fuse and drain into Lt atrium (this is different from anomalous unilateral single PV : only single PV)
  - Separated right middle lobe PV draining independantly from superior and inferior right PV.
  - Presence of > 2 pulmonary veins on the same side

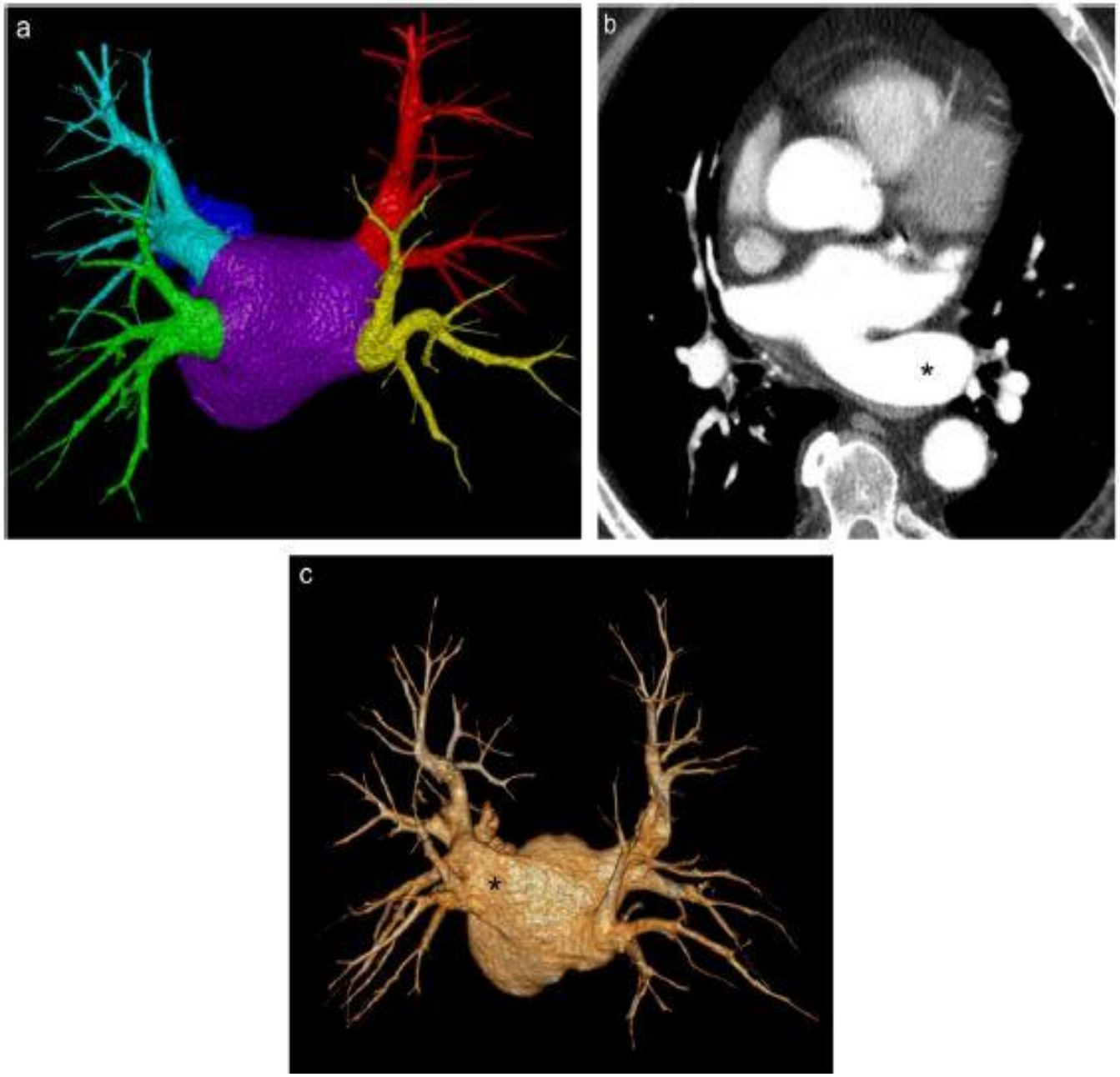


Fig. 11 Pulmonary venous variant. **a** Three-dimensional reconstruction of a contrast-enhanced CT shows normal pulmonary venous anatomy with two right and two left pulmonary veins draining into the left atrium. **b** Axial CT image showing the most common variant of pulmonary venous anatomy, a fusion of the left pulmonary veins prior to entry in the left atrium (*). **c** Three-dimensional reconstruction shows a fusion of the left superior and inferior pulmonary veins with a shared ostium (*)

Pulmonary Vein Abnormality

2. Anomalous pulmonary venous return

- PV flow drains into the systemic system producing left-to-right shunt
- Wide spectrum of tremendous variability
- Total anomalous pulmonary venous return(TAPVR) : all PV returns drains into the systemic system- incompatible with life, needs immediate corrective surgery
  - Classified into supracardiac, cardiac, infracardiac or mixed types depending on location of drainage.
- Partial APVR : only some of PV returns drains into the systemic system.
  - Severity of PAPVR depends degree of anomalous return, patient often asymptomatic.
- Larger defect indentified early in life, while smaller defect remaining asymptomatic(incidental findings in adulthood).
- Management : range from observation in mild cases to emergent surgery in severe cases.

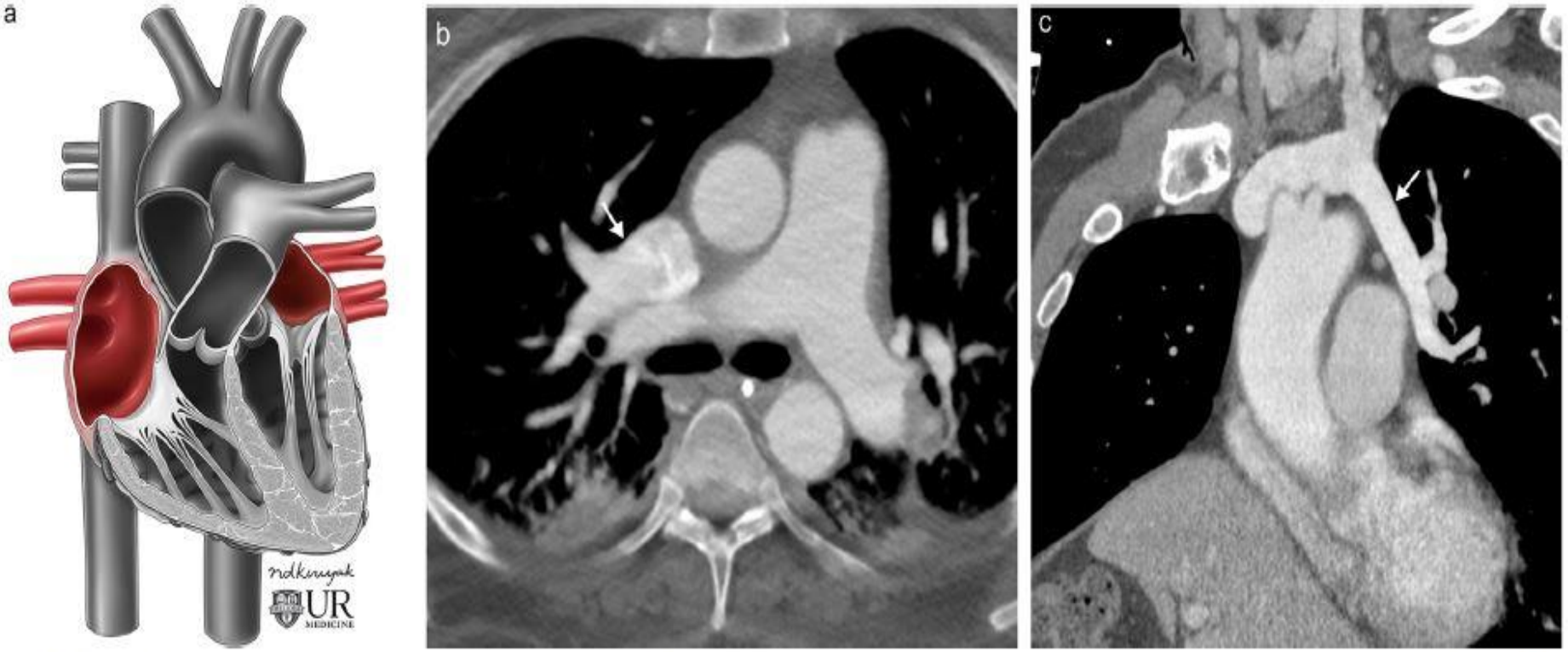


Fig. 12 Partial anomalous pulmonary venous return. **a** Conceptual illustration of partial anomalous pulmonary venous return. **b** Axial contrast-enhanced CT of an adult patient with partial anomalous venous return of the right upper lobe (*white arrow*) with blood flow returning to the superior vena cava. **c** Double oblique contrast-enhanced CT of an adult patient with partial anomalous venous return of the left upper lobe (*white arrow*) with blood flow returning to the left brachiocephalic vein

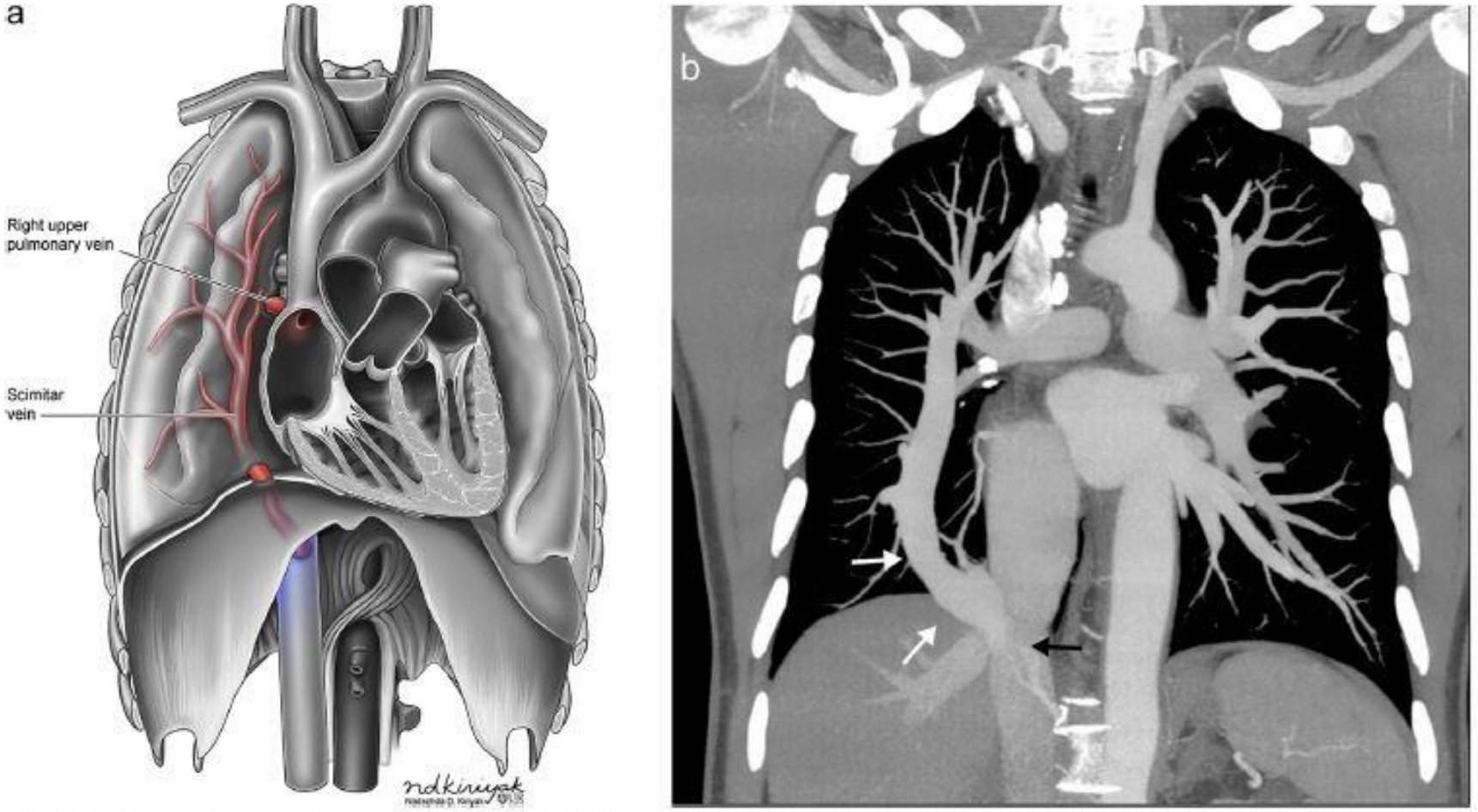


Fig. 13 Scimitar syndrome. a Conceptual illustration of scimitar syndrome. b Coronal contrast-enhanced CT of an adult patient with scimitar syndrome showing the entire right lung pulmonary venous return (*white arrows*) draining into the inferior vena cava (*black arrow*)

- Subtype of PAPVR : right sided PV drainage into the IVC (or other veins below), usually associated with right lung hypoplasia.

Pulmonary Vein Abnormality

3. Pulmonary vein varix/aneurysm

- Isolated segmental dilatation of PV aka varix or PV aneurysm
- Most frequently occur near the ostium
- CXR : pulmonary nodule or hilar lymphadenopathy
- Can also acquired secondary to cardiopulmonary disease eg. Chronic pulmonary hypertension
- Asymptomatic. If rupture →haemorrhage. Requiring surgery or coiling treatment.

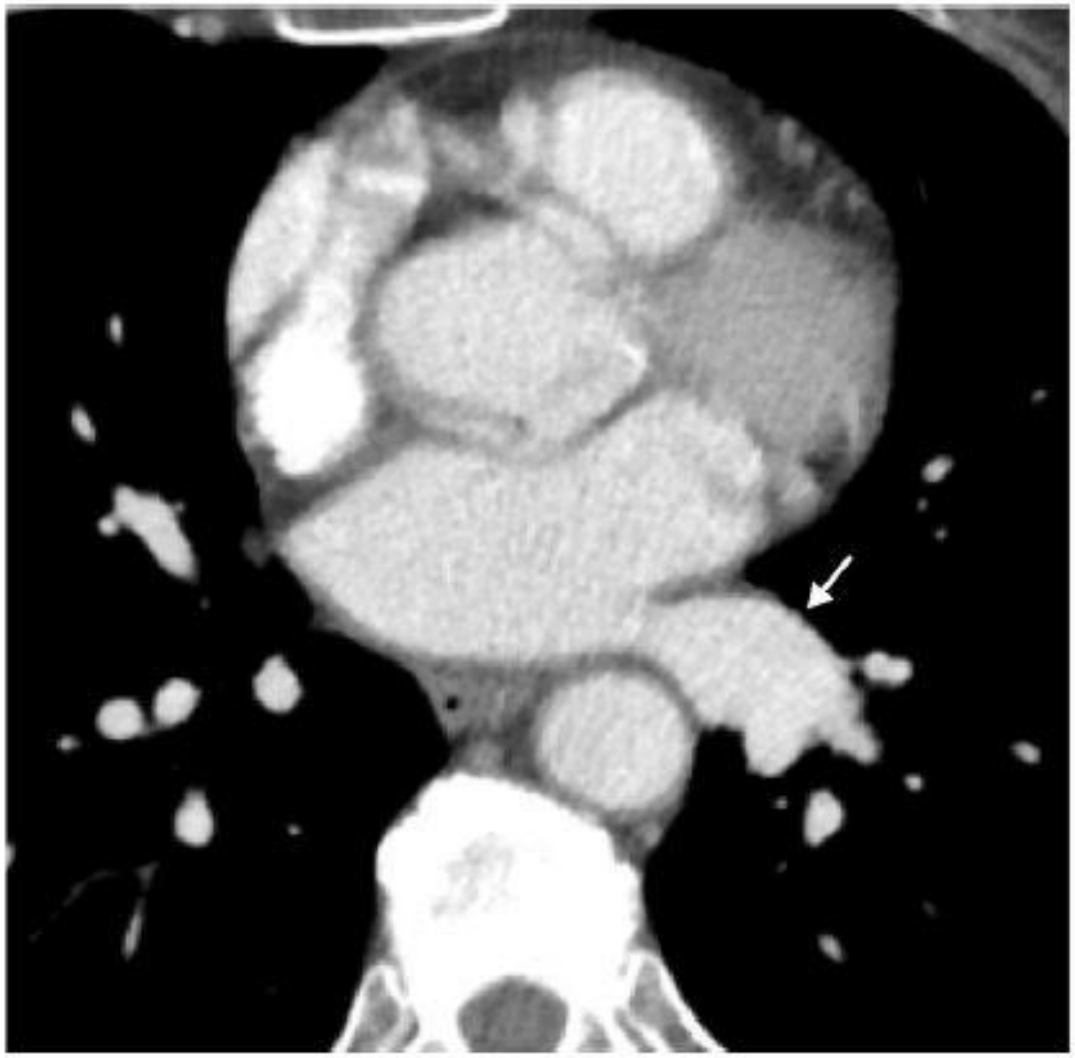


Fig. 14 Pulmonary vein varix. Axial contrast-enhanced CT image demonstrated a pulmonary vein varix (*white arrow*); note its proximity to the insertion in the left atrium. Varices can be saccular, tortuous, or confluent, with confluent being the most common

Pulmonary Vein Abnormality

4. Pulmonary vein congenital atresia/stenosis

- Failure of PV development
- Classically presented with recurrent pneumonia and/or hemoptysis in first few years of life
- Association with other congenital cardiac anomalies
- Divided into unilateral or bilateral PV atresia

Pulmonary Vein Abnormality

- ACQUIRED DISORDER OF PULMONARY VEIN

1. Pulmonary vein stenosis/obstruction

- Can be due to intrinsic or extrinsic processes.
- Majority cases due to neoplastic process. Other causes :
  - Radiofrequency ablation
  - Surgical complication
  - Fibrosing mediastinitis
  - Tuberculosis
  - Sarcoidosis
- These causes can cause PV thrombosis
- Clinical presentation is non specific
- Obstruction of PV can be detected on CT contrast imaging

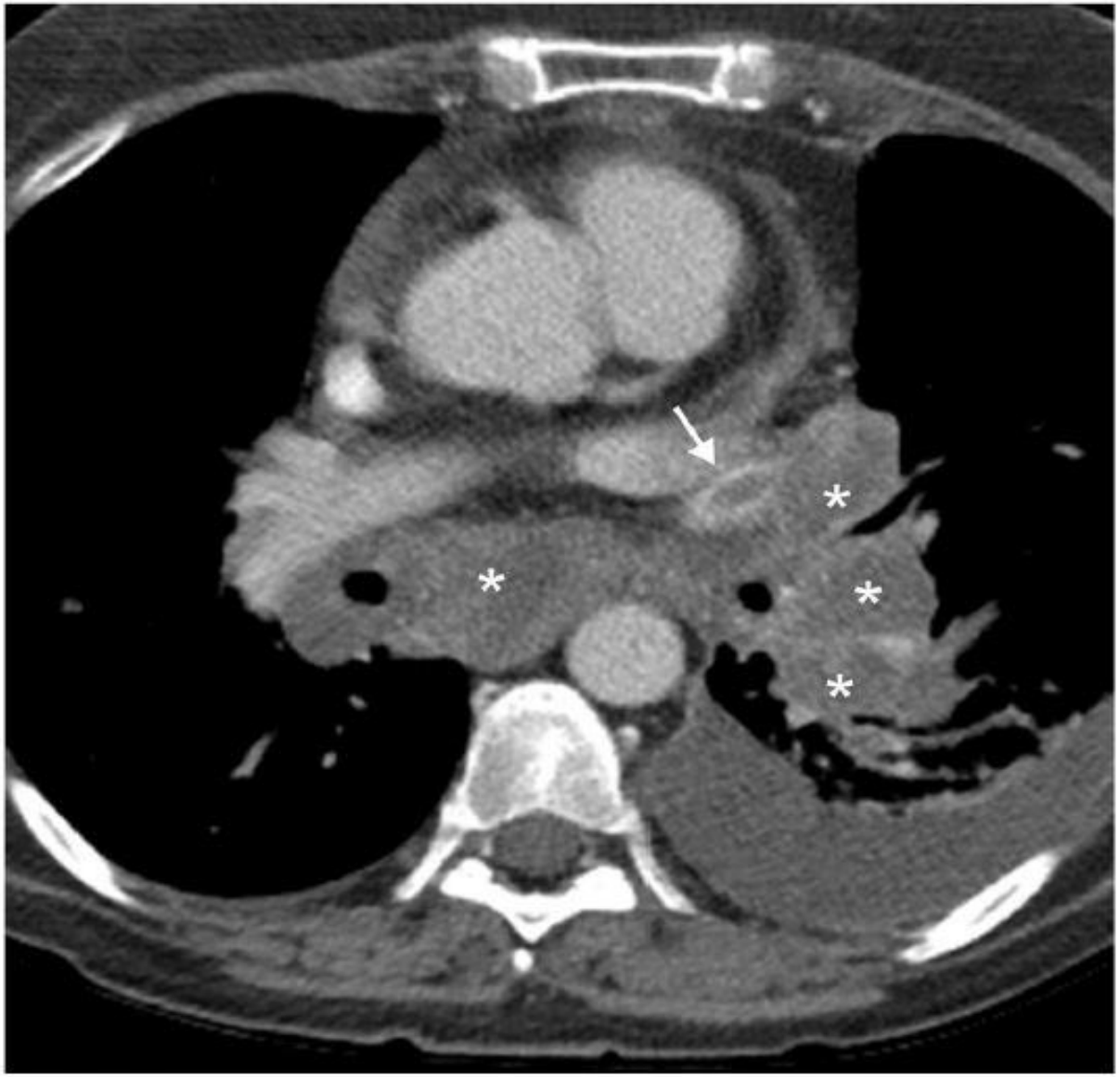


Fig. 15 Pulmonary venous thrombus. Axial contrast-enhanced CT image in a patient with non-small cell lung cancer demonstrating a filling defect within the left superior pulmonary vein (*white arrow*). Note extensive lymphadenopathy within the left hila and mediastinum (*) from metastatic involvement

Pulmonary Vein Abnormality

2. Pulmonary vein collateral pathways

- PV can be involved in a variety of collateral pathways, both congenital and acquired
- In cases of SVC obstruction, collaterals flow of PV can form, resulting in a right to left shunt

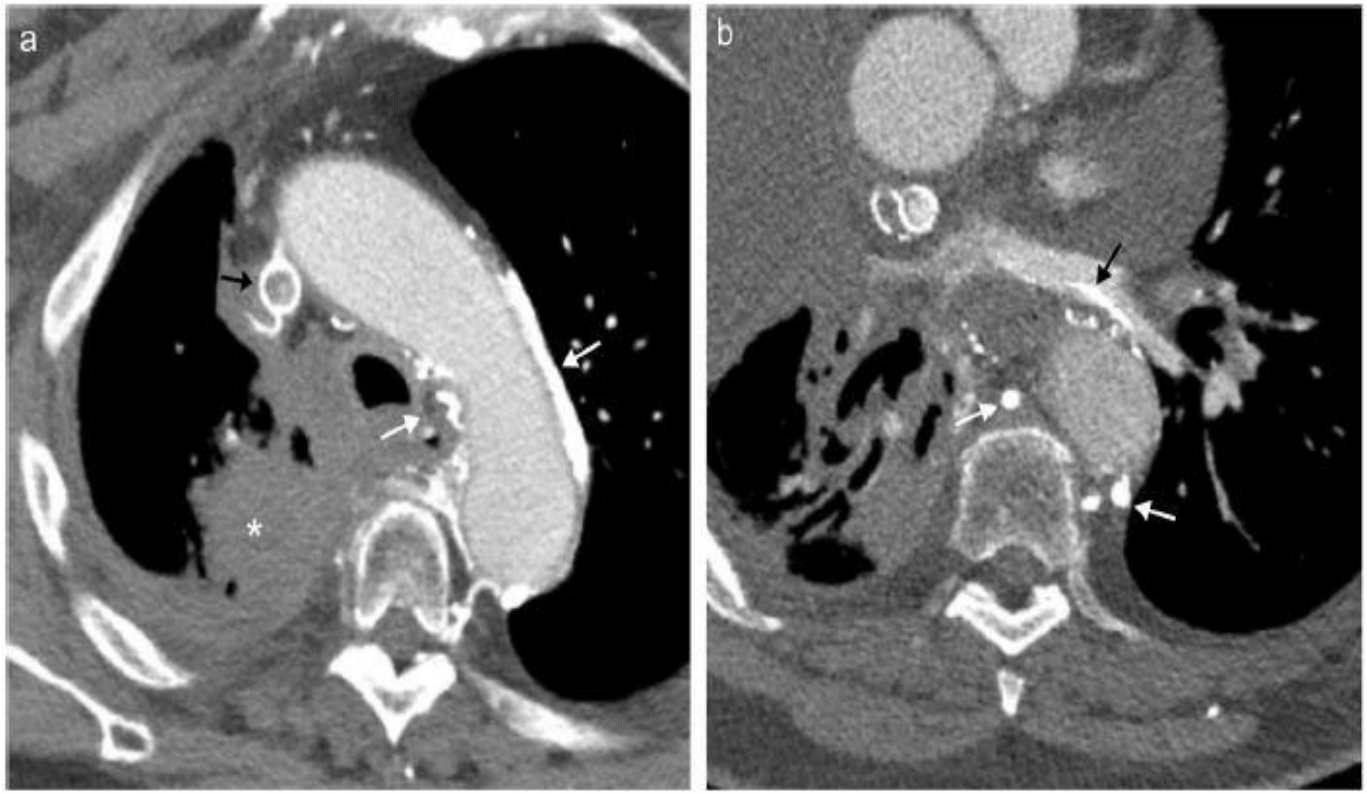


Fig. 16 Pulmonary venous collaterals. **a** Axial contrast-enhanced CT image demonstrates upper right lobe squamous cell carcinoma (*) with metastatic disease obstructing the superior vena cava and stent (*black arrow*). Note extensive collateral formation (*white arrows*). **b** Axial CT image showing high-density contrast flow into the pulmonary venous system (*black arrow*) secondary to collateral formation (*white arrows*) with a right-to-left shunt in the setting of superior vena cava obstruction

Pulmonary Vein Abnormality

3. Pulmonary venous hypertension

- Pressure equal or more than 15mmHg
- Most often in left ventricular failure or other cardiac causes
- A rare idiopathic cause of PV hypertension is known as pulmonary veno-occlusive disease : develop seconcary to occlusion or constriction of PV and venules
- Imaging may shows:
  - Fluid overload : pulmonary edema and pleural effusion
  - Mosaic attenuation of lung parenchyma
  - Enlargement of the central pulmonary arteries

Pulmonary Vein Abnormality

4. Pulmonary vein calcifications

- Typically associated with mitral valve disease and chronic renal failure
- May also seen in Atrial fibrillation patient

- Imaging shows:
  - Pulmonary vein calcifications
  - Dilated left atrium (may also calcified) described as “mould-like pattern”

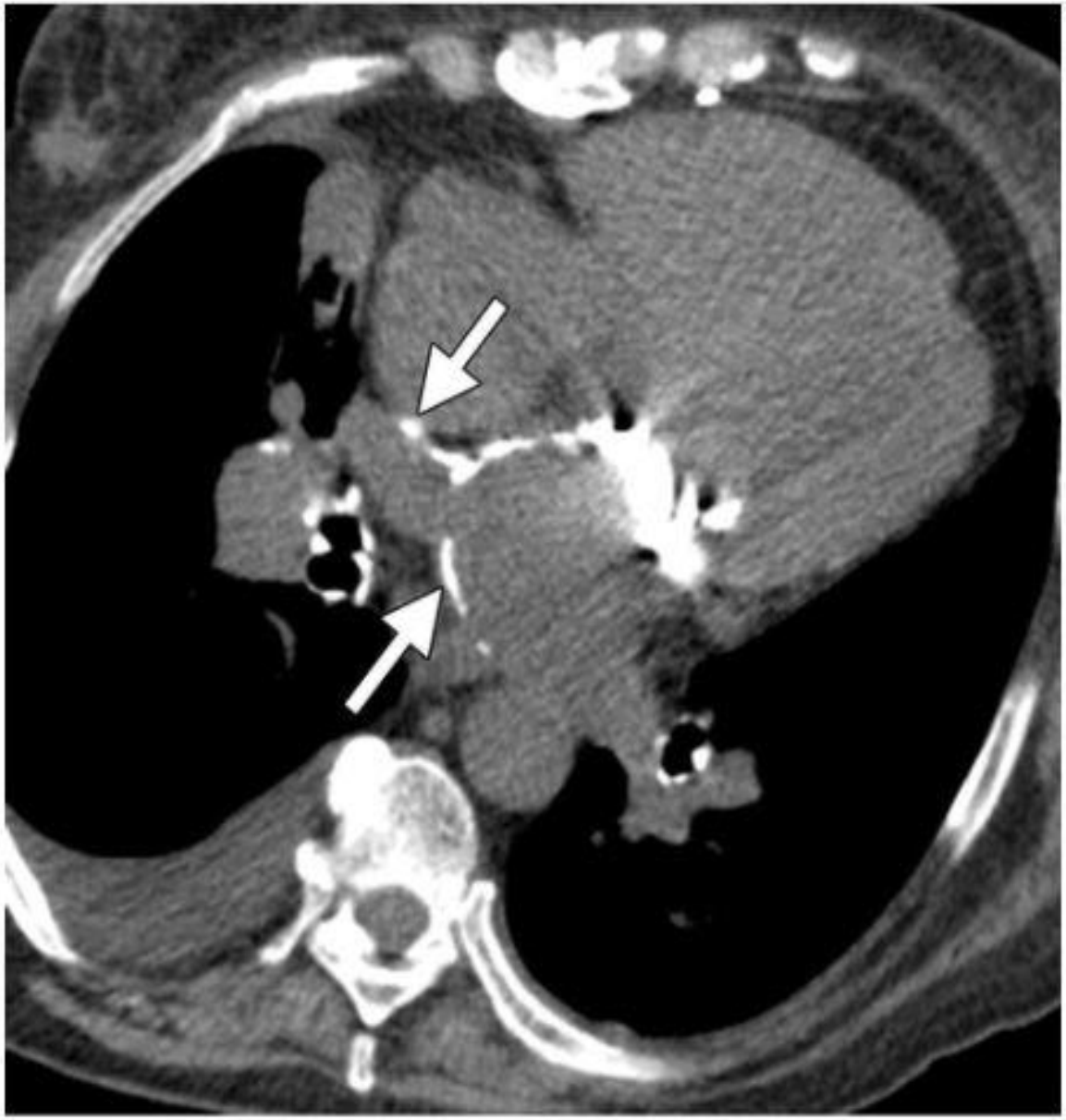


Figure 24 Rheumatic mitral valve stenosis treated with a mitral valve prosthesis in a 68-year-old woman who had atrial fibrillation. Axial nonenhanced multidetector CT image shows extensive wall calcifications (arrows) in the right superior pulmonary vein and left atrium.

OTHER PULMONARY VASCULAR ABNORMALITIES

1. Pulmonary arteriovenous malformations(AVM)

- Typically congenital, but only become clinically relevant in adulthood.
- Hereditary haemorrhagic telangiectasia : should considered if >1 AVM seen, as this condition associated with multi-organ AVM
- Can be acquired secondary to trauma, surgery for congenital heart disease, tuberculosis or shistosomiasis
- Imaging by CT is the best:
  - Well circumscribed non calcified nodule with large feeding artery and draining vein often at lower lobe
  - Calcified phleboliths may be present
  - Enhancement of feeding artery in early arterial phase
- Clinical findings depend on the size of malformation
- Lesions >2cm require treatment such as embolisation or surgically

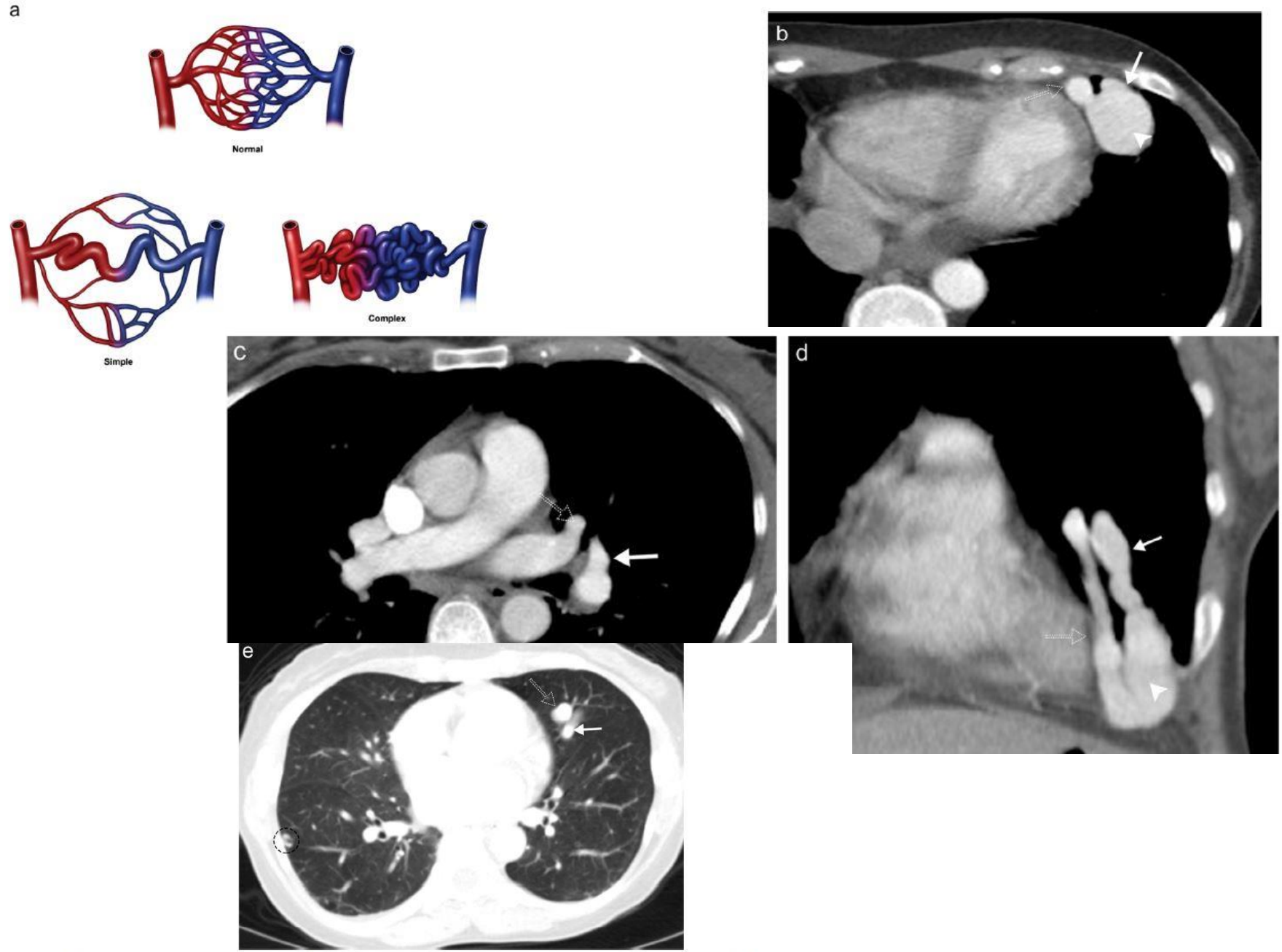


Fig. 17 Pulmonary arteriovenous malformation. **a** Conceptual illustration of simple versus complex arteriovenous malformation. Axial (**b**, **c**) and coronal (**d**) contrast-enhanced CT images demonstrate a large arteriovenous malformation of the left upper lobe (*arrowhead*) with a clearly identifiable feeding pulmonary artery (*solid arrow*) and draining pulmonary vein (*dotted arrow*). **e** Axial contrast-enhanced CT images of the same patient in lung windows reinforces the importance of fully evaluating the entire lungs as additional smaller AVMs (*circle*) were also identified in this patient leading to the diagnosis of hereditary haemorrhagic telangiectasia

OTHER PULMONARY VASCULAR ABNORMALITIES

2. Pulmonary sequestration

- Rare developmental abnormality : non-functional, aberrant lung tissue → no connection with tracheobronchial tree but still receiving systemic arterial flow(typically from aorta)
- Intra or Extralobar types : differ in their relationship with pleura, venous drainage and clinical presentation
- Intralobar (75% cases) typically present in childhood with recurrent pulmonary infection : adjacent to normal lung, without separate pleura, and the venous drain via pulmonary venous system (left-to-left shunt)
- Extralobar(aka accessory lung) : covered by own pleura, venous drainage into azygos system(left-to-right shunt) → typically diagnosed during infancy
  - Feeding difficulty or cyanosis episode
  - Associated with developmental abnormality eg. Scimitar syndrome or congenital heart disease

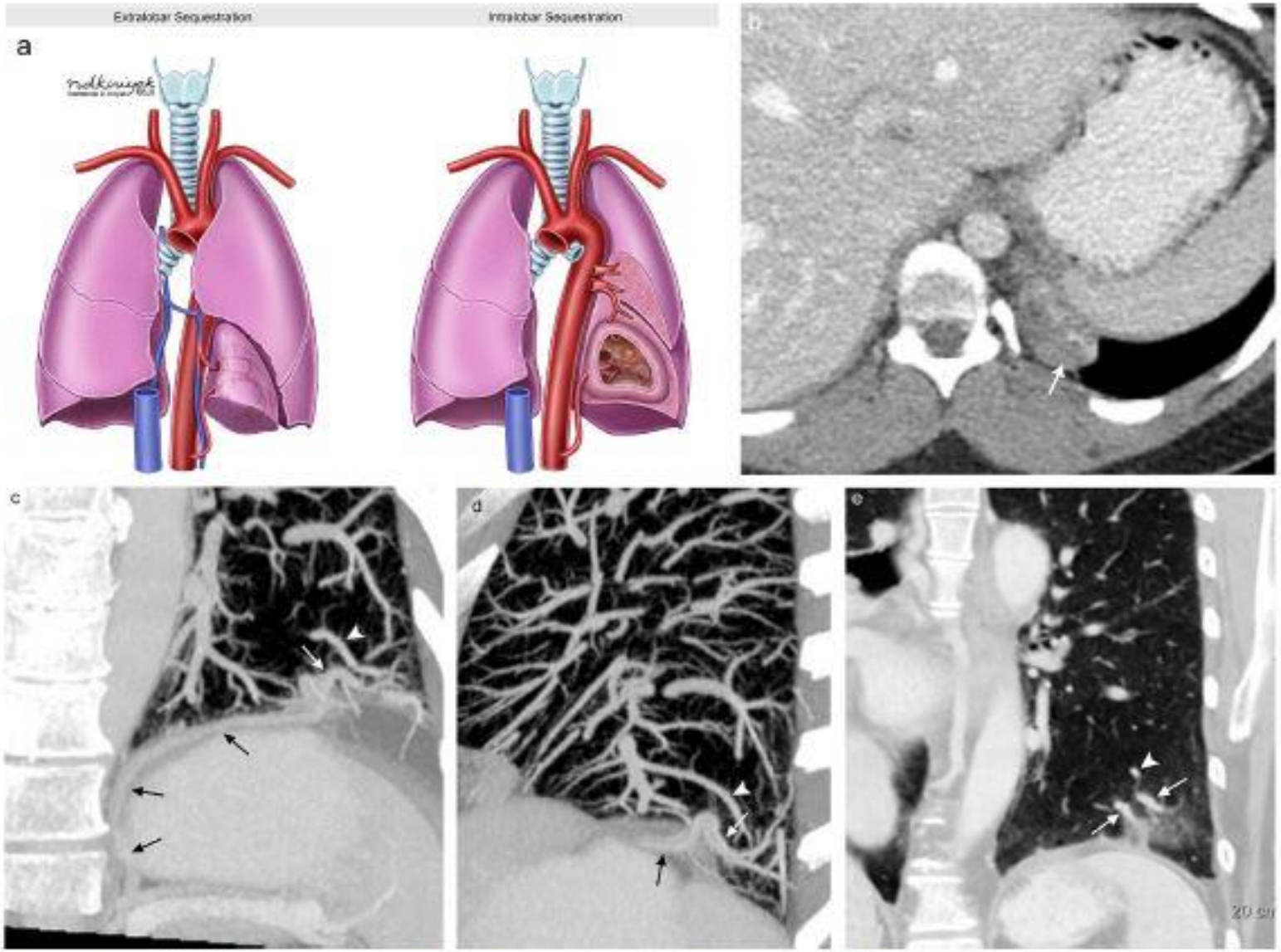


Fig. 18 Pulmonary sequestration. **a** Conceptual illustration of extralobar and intralobar sequestration. Note that extralobar sequestration involves venous drainage into the azygos system forming a left-to-right shunt while intralobar sequestration involves pulmonary venous drainage forming a left-to-left shunt. **b** Axial CT demonstrating an asymptomatic oval lesion (*white arrow*) in the left lower lobe consistent with extralobar sequestration in a 23-year-old woman. Blood supply is derived from the abdominal aorta with venous drainage occurring via the azygos system. Double oblique CT MIP images (**c**, **d**) and coronal CT MPR image (**e**) demonstrating an intralobar sequestration located within the pleura of the left lower lobe. Note the aberrant arterial supply to the lateral left lower lobe below the level of the diaphragm (*black arrows*) which extends through the diaphragm into the tissues of the left lower lobe (*white arrows*). Venous drainage of this intralobar sequestration is via the pulmonary venous system (*arrowheads*)

OTHER PULMONARY VASCULAR ABNORMALITIES

3. Bronchial artery abnormality

- Typically branch off from aorta but also can come from subclavian artery
- Classically 2 left and 1 right bronchial artery, measures <2mm
- Lot of anatomical variation, but typical variant shares common origin with intercostal artery known as intercostal-bronchial artery trunk (ICBAT)
- Not involve in alveolar gas exchange but if insufficient blood flow to the lung happen→undergo hypertrophy →form collateral pathway with pulmonary artery

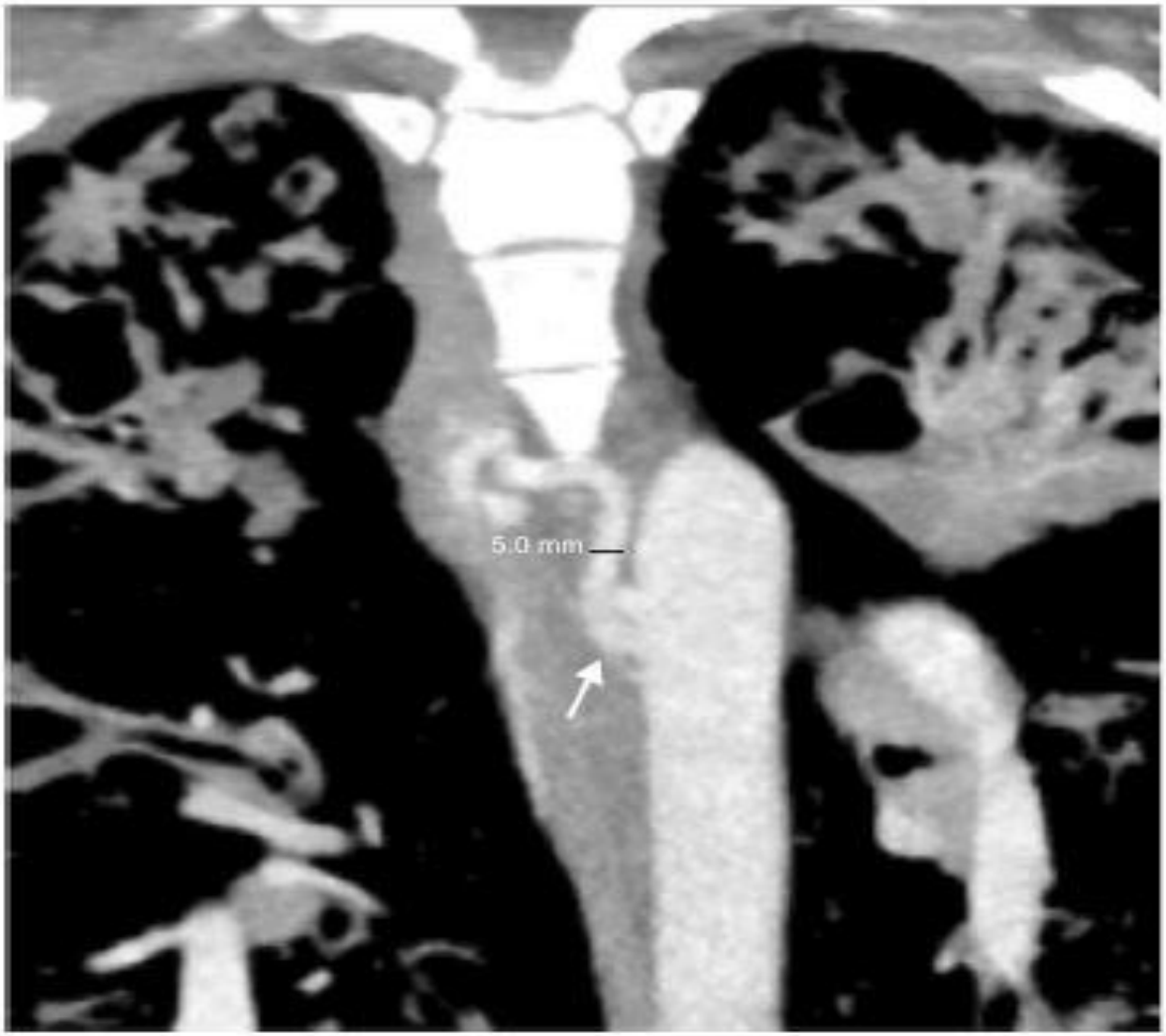


Fig. 19 Bronchial artery anatomy. Coronal CT of the chest shows a dilated right bronchial artery (5 mm) arising from the intercostal bronchial artery trunk (ICBAT) (*white arrow*) of the postero-lateral thoracic aorta at the level of T6. Please note that the normal bronchial artery is less than 2 mm in diameter. While ICBAT origin is the most typical of the right bronchial artery, the left bronchial arteries usually arise directly from the aorta

OTHER PULMONARY VASCULAR ABNORMALITIES

- Dilated bronchial artery (>2mm) : often sign of serious pathology
- Several causes of bronchial artery dilatation :
  - Congenital heart disease
  - Pulmonary artery obstruction
  - Inflammatory conditions of lung

- Patient with idiopathic pulmonary hypertension do not typically have bronchial artery dilatation : helpful clue in differential diagnosis

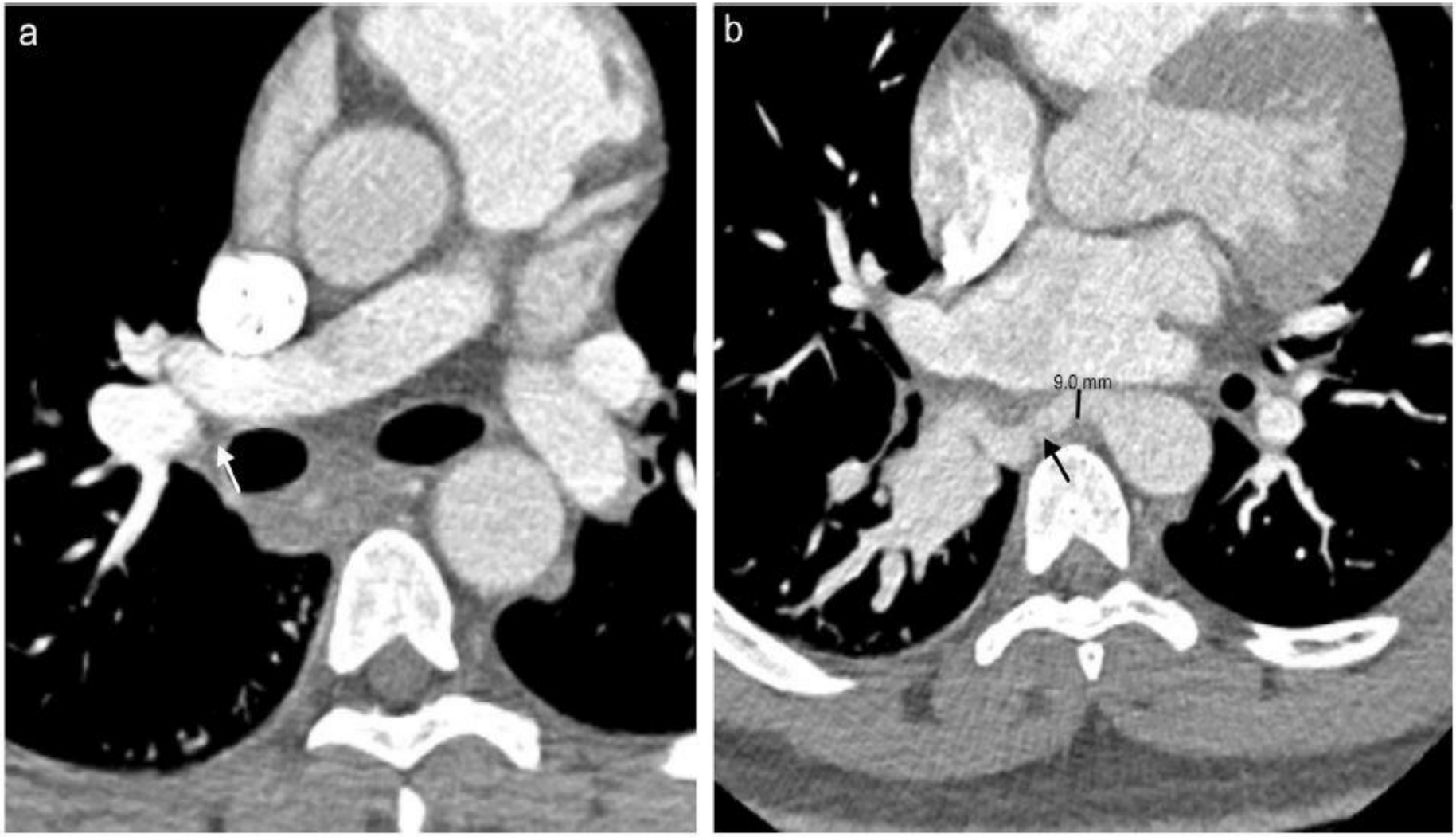


Fig. 20 Bronchial artery dilatation. **a** Axial CT image showing congenital absence of the right lower lobe pulmonary artery (*white arrow*). **b** Axial CT image showing 9.0 mm dilated bronchial artery supplying the right lower lobe (*black arrow*)

OTHER PULMONARY VASCULAR ABNORMALITIES

- Frequent complication of bronchial artery dilatation is hemoptysis
- Majority of hemoptysis that needs intervention are secondary to bronchial artery dilatation
- Massive hemoptysis : extremely high mortality rate
- CT angiography in aortic phase recommended for evaluation of bronchial artery prior to embolisation

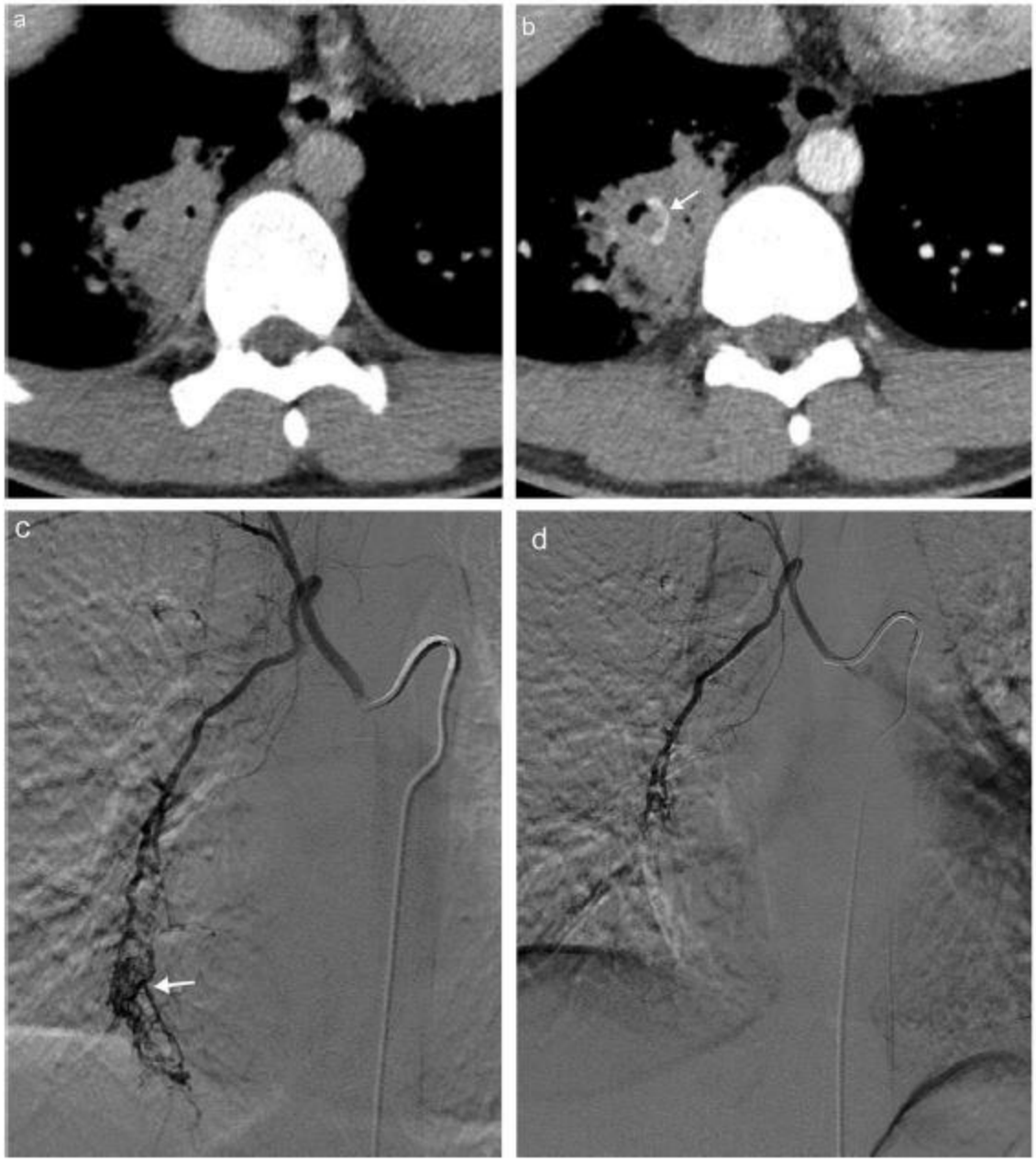


Fig. 21 Bronchial artery bleeding. Pre- (**a**) and post-contrast (**b**) axial images in a patient with pneumonia and new onset haemoptysis demonstrates a blush of contrast within the right lower lobe consolidation concerning for active bleeding (*arrow*). Bronchial arteriogram demonstrates a blush of contrast (*arrow*) confirming bleed pre embolisation (**c**) with resolution of the contrast blush post embolisation (**d**)

CONCLUSION

- Knowledge of these conditions and normal anatomy of pulmonary vasculature is the cornerstone for building a differential diagnosis tailored to patient's clinical presentation and imaging findings