Congenital malformations of the pulmonary vessels in adults.

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Learning objectives

- To know the radiologic manifestations of these malformations.
- To know the optimal imaging techniques and reconstructions to evaluate them.
- To know the clinical impact and possible treatment of these anomalies.

Background

Congenital lung malformations are rare. They can be classified into three categories: bronchopulmonary anomalies, isolated vascular anomalies, and combined pulmonary and vascular anomalies. These malformations are usually detected in the prenatal/neonatal period or in early childhood; however, some remain asymptomatic and are detected incidentally in adulthood.

These malformations have characteristic radiologic manifestations, although they can mimic other diseases and often cause diagnostic errors. Congenital lesions are normally characterized by CT or MRI with intravenous contrast agents and 3D reconstructions in different planes.

Findings and procedure details

1. PULMONARY ARTERIES:

- PROXIMAL INTERRUPTION OR ABSENCE OF THE MAIN PULMONARY ARTERY(Fig. 1 on page 13, Fig. 2 on page 13 and Fig. 3 on page 14):

  Incidence: 1/200,000 persons. This anomaly is characterized by the absence or anomalous termination of a pulmonary artery at the level of the hilum. The involved lung is irrigated through the collateral systemic circulation, mainly the bronchial arteries, although branches of the intercostal arteries, internal mammary artery, subclavian artery, and innominate artery can also contribute.

  The interrupted pulmonary branch is generally contralateral to the aortic arch; thus, the right pulmonary artery is more commonly affected, and in this case the anomaly is usually
asymptomatic. Interruption of the left pulmonary artery is often associated with a right aortic arch and other congenital cardiovascular defects like the tetralogy of Fallot.

**Clinical presentation:** This malformation can be asymptomatic, but most patients develop symptoms like recurrent infections, hemoptysis, and dyspnea. About 10% have hemoptysis due to rupture of hypertrophic collateral arteries, and this is the most important finding for the prognosis in 19% to 25% of patients.

**Plain-film X-rays:**

- Various degrees of pulmonary hypoplasia, elevation of the ipsilateral diaphragm, and displacement of the heart and mediastinum toward the affected side.
- Compensatory hyperinsufflation of the contralateral lung, which herniates toward the affected hemithorax.
- Subpleural reticular opacities due to collateral systemic vessels that supply the affected lung.
- Notches in the ipsilateral ribs, when the collateral vessels depend on the intercostal arteries.

**Multidetector CT:**

- Is the best way to visualize the abnormal pulmonary artery, which can be completely absent or can terminate 1 cm from its origin.
- Shows the hypertrophied systemic arteries.
- Shows associated anomalies: serrated pleural thickening and parenchymatous bands beneath the pleura representing anastomoses of the transpleural collateral arteries with the distal pulmonary arteries.

**Treatment:** In children and young adults it is sometimes possible to revascularize the pulmonary artery with reimplantation or shunting of the affected segment. When the malformation is detected in adults, the treatment is symptomatic. The systemic arteries can be embolized in cases of recurrent hemoptysis or the lung can be resected to definitively stop persistent bleeding.

**ANOMALOUS ORIGIN OF THE LEFT PULMONARY ARTERY (PULMONARY ARTERY SLING) (Fig. 4 on page 15 and Fig. 5 on page 16):**

Incidence: 59/1,000,000 persons. In this malformation, the left pulmonary artery arises from the posterior aspect of the extrapericardial segment of the right pulmonary artery and courses between the trachea and esophagus to reach the left hilum, forming a sling around the distal trachea and the proximal main right bronchus.
**Clinical presentation:** Patients can be classified into two groups: those with a normal tracheobronchial pattern, including asymptomatic adults, and those with one or more malformations of the tracheobronchial tree (e.g., stenosis of a long segment of the trachea or absence of the pars membranacea) and/or cardiovascular malformations; this second group has high morbidity and mortality during childhood. Symptomatic patients normally have stridor, wheezing, and recurrent lung infections.

**Plain-film X-rays:**

- AP view: opacity measuring approximately 2 cm in diameter in the tracheobronchial angle.
- Lateral view: rounded opacity between the trachea and esophagus.

**Multidetector CT:**

- Shows the anomalous origin and course of the artery and makes it easier to evaluate the airway through MPR and 3D reconstruction techniques.

**Treatment:** depends on the associated anomalies, particularly those involving the tracheobronchial tree.

**IDIOPATHIC DILATION OF THE PULMONARY TRUNK (Fig. 6 on page 17):**

This uncommon anomaly is characterized by abnormal widening of the pulmonary trunk, with or without widening of the main pulmonary arteries. This is a benign condition that does not progress, and affected individuals are asymptomatic. The diagnosis is reached by ruling out lung and heart disease and confirming normal pressure in the right ventricle and pulmonary artery. The main differential diagnoses include congenital stenosis of the pulmonary valve and pulmonary arterial hypertension.

**Plain-film X-rays:**

- Enlargement of the pulmonary trunk, which manifests as a convex opacity that mimics a mediastinal mass.

**Multidetector CT:**

- Increased diameter of the main trunk of the pulmonary artery (>29 mm); no increase in intraparenchymal arteries is seen.

**PULMONARY VALVE STENOSIS (Fig. 7 on page 18):**
This condition is congenital in 95% of cases and is found in about 10% of all patients with congenital heart defects. Pulmonary valve stenosis is by far the most common cause of reduced right outflow (90% of cases), followed by subvalvular and supravalvular stenoses. It is characterized by fusion of the leaflets of the valves at the commissures.

**Clinical presentation:** The symptoms depend on the severity of the stenosis. Patients with mild stenosis are asymptomatic; those with moderate or severe stenosis can have signs and symptoms of systemic venous congestion similar to that seen in congestive heart failure.

**Plain-film X-rays:**
- Enlargement of the pulmonary trunk and of the left pulmonary artery (due to the direction of the jet in the regurgitation).
- Because the size of the right pulmonary artery is normal, the hila can be asymmetrical.

**Multidetector CT:**
- Poststenotic dilation of the pulmonary trunk and of the left pulmonary artery.
- Occasionally, dilation of the right ventricle.
- The leaflets of the pulmonary valves can be thickened and calcified.

**MRI:**
- Useful for evaluating the morphology of the pulmonary valve and characterizing the direction, size, and range of the abnormal flow associated with pulmonary stenosis.
- Thickening and fusion of the leaflets of the valve and narrowing of the opening of the valve can be observed.
- Additional morphological abnormalities can be identified (e.g., bulging and reduced movement of the pulmonary valve and hypertrophy and thickening of the right ventricle).

**Treatment:** depends on the hemodynamic consequences; when no other heart defects are present, percutaneous balloon valvuloplasty can be done. Occasionally, pulmonary valve replacement is necessary.

2. PULMONARY VEINS:

- **STENOSIS/ ATRESIA OF THE PULMONARY VEINS:**
Stenosis of the pulmonary veins is usually acquired (reconstructive surgery, pulmonary venous ablation). Congenital stenosis is rare and seems to be caused by uncontrolled proliferation of fibroblasts, which results in thickening and stenosis of the vein.

**Clinical presentation:** affected patients have symptoms similar to those with pulmonary edema, including cyanosis, dyspnea, pulmonary hypertension, recurrent respiratory infections, or hemoptysis.

**Plain-film X-rays:**
- Changes due to reduced venous drainage in the lungs and resulting pulmonary vein hypertension.
- Reduced volume of the affected lung.
- Thickening of the interlobar septa and an increase in the diffuse reticular opacities in the entire affected lung, compatible with pulmonary edema.

**Multidetector CT:**
- The involved segment of the pulmonary vein appears stenosed with thickened walls, generally where the vein empties into the left atrium.
- In advanced stages of the disease, involvement can extend to the most distal portions of the pulmonary vein.
- Associated findings can include ipsilateral pleural effusion, thickening of the interlobar septa, and ground-glass opacities due to unilateral pulmonary edema.

**Treatment:** Stenoses of short segments can be treated with balloon dilation and stenting; however, stenoses of long segments require lung transplantation.

· **PULMONARY VARICES** *(Fig. 8 on page 19)*:

Pulmonary varices are characterized by aneurysmatic dilation of a segment of the pulmonary vein, generally where it joins the left atrium. Most cases are congenital, although pulmonary varices can be acquired in the context of mitral valve disease or pulmonary hypertension.

**Clinical presentation:** Most patients are asymptomatic and the varices are detected incidentally. On rare occasions, patients can have complications such as hemoptyisis or thrombosis.

**Plain-film X-rays:**
• Well-defined nodular or tubular opacities in the walls near the margins of the cardiomeediastinal silhouette that can vary with Valsalva maneuvers; the findings can be mistaken for infectious or neoplastic processes.

**Multidetector CT** (with contrast administration), considered the imaging technique of choice:

• Aneurysmatic dilation of a segment of the pulmonary vein.
• Absence of an afferent artery differentiates pulmonary varices from an arteriovenous malformation.

**Treatment:** Surgery is only indicated in cases with recurrent hemoptysis or large increases in the caliber of the vein.

**PARTIAL ANOMALOUS PULMONARY VENOUS RETURN (PAPVR): HYPOGENETIC LUNG SYNDROME AND OTHER ANOMALOUS VENOUS DRAINAGE:**

PAPVR can be divided into two large groups: (a) hypogenetic lung syndrome or Scimitar syndrome, which involves a malformation of the right lung and an anomalous pulmonary vein that is easily identified in plain-film X-rays, and (b) other, usually asymptomatic, types of PAPVR that are not associated with lung abnormalities.

**HYPOGENETIC LUNG SYNDROME (SCIMITAR SYNDROME)** *(Fig. 9 on page 20 and Fig. 10 on page 21):*

This is a type of PAPVR in which the anomalous pulmonary vein is the vein that drains all or part of the right lung, emptying into the inferior vena cava either above or below the diaphragm, although occasionally it can empty into the hepatic vein, portal vein, azygos vein, coronary sinus, or right atrium. It is generally associated with various degrees of right lung hypoplasia, with a hypoplastic or aplastic pulmonary artery and dextrocardia. The anomalous pulmonary drainage results in a left-right shunt. The right lung usually has abnormal lobulation (often only two lobes are present), and the bronchographic pattern mimics that of the left lung. About 25% of these patients have congenital heart defects, the most common being interatrial communication. Other associated anomalies include bronchogenic cyst, pulmonary sequestration, horseshoe lung, and diaphragmatic anomalies.

**Clinical presentation:** In 10% of cases, patients are asymptomatic and the anomaly is discovered incidentally on plain-film chest X-rays. The symptoms depend on the magnitude of the associated left-right shunt and possible development of pulmonary hypertension.
Plain-film X-rays:
- Can diagnose the condition when an anomalous pulmonary vein is seen as a vertically oriented curved line (resembling a scimitar) in the right hemithorax.
- Loss of volume in the right lung and hyperinsufflation of the contralateral lung.

Multidetector CT:
- Enables evaluation of the course of the anomalous vein and the place where it empties.
- Hypoplasia of the right lung; abnormal lobulation and bronchial tree.
- Other findings include a hypoplastic or aplastic right pulmonary artery, hypertrophy of the systemic circulation, absence of the inferior vena cava, accessory diaphragm, horseshoe lung, and congenital heart defects.

Treatment: Except when necessary for associated heart defects, treatment is rarely indicated in adults. Treatment usually involves reimplantation of the anomalous pulmonary vein into the left atrium or interrupting the systemic arterial supply to the lung.

ANOMALOUS PULMONARY VENOUS RETURN, ISOLATED OR ASSOCIATED WITH HEART DISEASE (Fig. 11 on page 22, Fig. 12 on page 23 and Fig. 13 on page 24):

PAPVR can be associated with congenital heart disease; however, more frequently it is an isolated finding. This occurs when one or more pulmonary veins drain directly into the systemic circulation (normally ipsilateral) causing a left-right shunt. Right PAPVR usually drains into the azygos vein, superior vena cava, right atrium, inferior vena cava, or suprahepatic veins; left PAPVR drains into the vertical vein, hemiazygos system, or coronary sinus.

Types:

1. Partial: when one or more veins, but not all, are connected to the systemic circulation. In 90% of cases, this drainage is associated with sinus venosus atrial septal defects.
   - PAPVR of the right upper lobe draining into the caudal portion of the superior vena cava just above the atrium.
   - PAPVR of the left upper lobe through the vertical vein in the left innominate vein.

2. Total: predominantly seen in children. There is no connection between the pulmonary veins and the left atrium. Comprises a wide spectrum of heart defects.
**Clinical presentation:** most patients with isolated PAPVR are asymptomatic.

**Plain-film X-rays:** findings can be normal, although if the left-right shunt is pronounced, we can see increases in the pulmonary circulation and in the right chambers of the heart.

**Multidetector CT:** shows the pulmonary vessels in great detail, including the course of the anomalous pulmonary veins.

**MEANDERING PULMONARY VEIN** *(Fig. 14 on page 25):*

This condition consists of a pulmonary vein with an abnormal sinuous path but normal drainage into the left atrium, so no shunt is present.

**Clinical presentation:** patients are asymptomatic, and the condition is nearly always discovered incidentally on a plain-film X-ray or CT of the chest.

**Plain-film X-rays:**

- Abnormalities in the caliber and number of pulmonary veins are nearly always present.
- Arcuate or tortuous tubular opacities.
- The course of the vein often resembles the handle of a bucket.

**Multidetector CT:**

- The entire course of the anomalous vein can be seen, including where it empties into the left atrium.
- Associated anomalies may be detected (e.g., pulmonary artery hypoplasia, a dilated or tortuous pulmonary artery, bronchopulmonary malformations, abnormal systemic arteries, or PAPVR in a different lung region).

**Treatment:** the management depends on patients' symptoms and associated pulmonary anomalies.

3. **COMBINED ARTERIAL AND VENOUS ANOMALIES.**

**ARTERIOVENOUS MALFORMATIONS (AVM)** *(Fig. 15 on page 26 and Fig. 16 on page 27):*
An AVM results from a focal defect in the development of the lung's capillary network. This defect results in a direct communication between a branch of the pulmonary artery and its adjacent pulmonary vein. Most are congenital, associated with the autosomal dominant Rendu-Osler-Weber syndrome. AVMs are termed "simple" when the feeding vessels come from the same segmental artery and "complex" when they come from various segmental arteries.

**Clinical presentation:** AVMs are found in the following situations:

- Incidentally in a routine chest X-ray or CT study.
- Secondary to complications: hypoxia and cyanosis (if there is a significant shunt); septic or thrombotic paradoxical emboli; and rupture in a bronchus (hemoptysis) or in the pleural cavity (hemothorax).
- In the workup for Rendu-Osler-Weber syndrome (clinical triad of epistaxis, telangiectasias, and family history).

**Plain-film X-rays:**

- One or more well-circumscribed nodular or serpiginous structures with the density of soft tissue, predominantly in the lower lobes (50% - 70%).
- Occasionally, the feeding artery, the dilated draining vein, or both can be seen as a curvilinear opacity arising from the lesion and coursing toward the mediastinum.

**Multidetector CT** (technique of choice) provides information that is fundamental prior to embolization:

- It enables the identification of the AVM and its feeding and draining vessels, making it possible to determine whether it is simple or complex.
- The number of AVMs and the size of the feeding arteries and draining veins are important for planning treatment.
- MPR and 3D reconstructions are especially useful in cases with complex AVMs.

**Treatment** is usually done only when the diameter of the feeding arteries is greater than 3 mm in diameter. Endovascular embolization with coils or balloon occlusion are the techniques of choice.

4. **SYSTEMIC ARTERIES.**

· **BRONCHOPULMONARY SEQUESTRATION** (Fig. 17 on page 28):
This anomaly consists of a segment of nonfunctional lung parenchyma that is separated from the tracheobronchial tree and has systemic blood supply. The systemic supply usually comes from the descending thoracic aorta; however, in 20% of cases it comes from the superior abdominal aorta. This anomaly can be intralobar or extralobar: intralobar sequestrations are located in the visceral pleura of the lung, whereas extralobar sequestrations are separated from the normal lung and enveloped in their own pleural membrane.

Intralobar sequestration is more common than extralobar sequestration (3:1). In 95% of cases intralobar sequestrations are drained by the pulmonary veins, resulting in a left-left shunt.

Extralobar sequestration is often associated with other congenital malformations (diaphragmatic hernia, heart defects, or congenital malformation of the pulmonary airway); venous drainage takes place through the azygos vein, hemiazygos vein, and inferior vena cava, resulting in a left-right shunt. For this reason, extralobar sequestrations are usually diagnosed in the neonatal period, whereas intralobar sequestrations are usually discovered in adults.

**Clinical presentation:** Intralobar sequestrations are often asymptomatic and discovered incidentally on chest X-rays or CT. The most common clinical manifestations are recurrent infections and hemoptysis due to bleeding from the systemic artery, which is often malformed. Since extralobar sequestrations are not connected to the lung, they rarely become infected.

**Plain-film X-rays:**

- A homogeneous focal lung mass in the posterior basal segment of a lower lobe, most often the left, contiguous with the hemidiaphragm.
- Other, less common findings include a focal radiolucent area, cystic mass, or prominent vessels in the lower lobe.
- The aberrant systemic artery can be evident.

**Multidetector CT:**

- **Intralobar sequestration:**
  - Focal areas of radiolucency or complex cystic lesion with internal cavitation that might have air-fluid levels.
  - Other, less common findings include cysts and nodules, solid masses, mucoid impaction, and calcifications.
  - Anomalous arterial vessel and venous drainage, typically to the inferior pulmonary vein.
- Areas of emphysema or air trapping in the lung adjacent to the lesion, in both intralobar and extralobar sequestration.

- **Extralobar sequestration:**
  - Homogeneous opacity or mass with well-defined margins.
  - Occasionally, cystic areas.
  - Anomalous arterial vessel and venous drainage into the systemic circulation via the azygos vein, hemiazygos vein, and inferior vena cava.

  **Treatment:** surgical resection (risk of recurrent infection /hemoptysis). As an alternative, it is also possible to embolize the anomalous feeding vessel.

- **SYSTEMIC IRRIGATION OF HEALTHY LUNG TISSUE** ([Fig. 18 on page 29]):

  This condition consists of a systemic artery that feeds normal lung tissue without associated alterations.

  **Clinical presentation:** patients can be asymptomatic, or they can have hemoptysis or hemothorax (due to rupture of the anomalous artery) or heart failure secondary to a left-left shunt between the anomalous artery and the pulmonary venous return.

  **Plain-film X-rays:** normal findings, except in cases in which the anomalous artery is very long or aneurysmatic.

  **Multidetector CT:**
  - The systemic artery arises from the thoracic or abdominal aorta and courses to the lower lobe through the inferior pulmonary ligament. It normally follows an ascending then descending course until it divides into multiple branches in the basal segments, adopting the trajectory and distribution of the pulmonary arteries.
  - The anomalous systemic artery can have atheromatous lesions such as calcified plaques and dilations or stenosis, even in young adults.
  - Other findings include bronchial dilation in the posterior and basal segment and ground-glass appearance in the parenchyma fed by the anomalous vessel, due to increased perfusion or hemoptysis.

  **Treatment:** Nowadays, embolization of the anomalous vessel with coils. In the past, surgical ligation of the anomalous artery and resection of the involved lung segment.
Figure 1. Asymptomatic unilateral proximal interruption of the right pulmonary artery in a 48-year-old man. (a) The posteroanterior chest radiograph shows a small hemithorax, mediastinal shift (↓), absence of the right pulmonary artery shadow (🪆), and linear opacities that correspond to systemic collateral vessels (➡) along the pleura and within the lung. This patient did not manifest pulmonary hypertension.

Fig. 1

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Figure 2. **Unilateral proximal interruption of the right pulmonary artery** in a 52-year-old woman with progressive shortness of breath and hemoptysis. (a) Contrast material–enhanced CT scan shows only the proximal portion of the right pulmonary artery (▲) and enlargement of the main and left pulmonary arteries that indicates pulmonary hypertension. (b) Contrast-enhanced CT scan at the level of the upper lobes shows serrated thickening of the right pleura because of enlarged intercostal collateral vessels (▲). (c) CT scan obtained with a lung window setting shows multiple linear opacities perpendicular to the pleural surface that correspond to transpleural systemic vessels (▲).

**Fig. 2**

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Figure 3. **Unilateral absence of the left pulmonary artery** in a 11-year-old girl with other associated malformations (a) Contrast-enhanced CT scan shows the absence of the left pulmonary artery (▲) and a right aortic arch (▲). (b) Contrast-enhanced CT scan coronal images shows systemic left lung irrigation from the thoracic and abdominal aorta (▲). (c) CT scan obtained with a lung window setting shows left pulmonary hypoplasia, leftward displacement of the heart and multiple linear opacities perpendicular to the pleural surface that correspond to transpleural systemic vessels (▲).

Fig. 3

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Figure 4. Diagram shows the anomalous origin of a left pulmonary artery (P.A.) that arises from the posterior aspect of the right pulmonary artery and reaches the left hilum by passing between the trachea and the esophagus.

Fig. 4

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Figure 5. Anomalous origin of the left pulmonary artery in a 60-year-old asymptomatic woman. (a) Posteroanterior chest radiograph shows an anomalous right paratracheal border (▲). (b, c) Unenhanced CT scan (b) and MR angiogram (c) at the level of the pulmonary trunk show the abnormal course of the left pulmonary artery (▲ in b) between the lower portion of the trachea and the esophagus (*) in b. (Fig 5b and 5c reprinted, with permission, from reference 12.)

Fig. 5

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Figure 6. Idiopathic dilatation of the pulmonary trunk in a 55-year-old asymptomatic woman. (a) Contrast-enhanced CT scan shows abnormal enlargement of the main pulmonary trunk, with mild dilatation of the right and left pulmonary arteries. (b) CT scan obtained with a lung window setting at the same level as b shows normal vessels and parenchyma. (c) Posteroanterior chest radiograph shows an abnormal bulge in the left mediastinal border (Δ), a feature suggestive of a mediastinal mass identical to that observed on radiographs obtained 6 years earlier (not shown).

Fig. 6

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Figure 7. **Pulmonic stenosis** in a 74-year-old woman. (a) PA chest radiograph shows an abnormal bulge in the left mediastinal border (△) that corresponds with the pulmonary trunk and the left pulmonary artery dilatation. (b) Contrast-enhanced axial CT shows thickening of pulmonic valve leaflets (★). (c) Sagittal image and (d) 3D reformations showing thickening of pulmonary valve leaflets (★) and pulmonary trunk and left pulmonary artery enlargement (★).
Figure 8. Pulmonary varix in a 21-year-old man. (a) Enhanced axial CT image shows an enlarged superior left pulmonary vein (▲) with an anomalous path draining in the left atrium. (b) Coronal MPR shows the atypical left superior pulmonary vein (▲). (c and d) CT scan obtained with a lung window setting shows important air trapping in inferior left lobe and lingula due to the compression by the anomalous vein on lingula’s bronchus (▲).

Fig. 8

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Figure 9. A 34-year-old woman with scimitar syndrome. Frontal chest radiograph shows multiple curvilinear tubular opacities (►) in right pulmonary lobe.

Fig. 9

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Figure 10. Same patient than figure 9. (a) Axial contrast enhanced CT demonstrates a large lower lobe pulmonary vein (▲) draining into the IVC and hypogenetic right lung. (b) Coronal MIP images demonstrates right lower lobe pulmonary vein (▲) draining into IVC (●). (c) CT scan obtained with a lung window setting shows right bronchial anomalous segmentation and a cystic dilatation of a right lower lobe segmentary bronchus (●).

Fig. 10

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Figure 11. 45 years-old man with PAPVR draining into SVC. (a) Axial CT contrast-enhanced image shows superior lobe vein (▲) draining into SVC (★). (b) MPR image in an axial-oblique plane demonstrates the termination of the abnormal vein in the lower portion of the SVC (▲).

Fig. 11

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**Figure 12.** 71 years-old woman with bilateral PAPVR draining into SVC and left brachiocephalic trunk. (a and b) Axial CT contrast-enhanced image shows right superior vein (▲) and the middle lobe vein (▲) draining into SVC (★). (c) MIP image in an axial-oblique plane demonstrates the drainage of the abnormal upper left lobe vein (▲) in the left brachiocephalic trunk through the vertical vein (†).

**Fig. 12**

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Figure 13. 83 years-old woman with PAPVR draining into SVC associated with interatrial communication. (a and b) Axial and coronal CT contrast-enhanced image shows upper lobe vein (▲) draining into SVC (★), associated with pulmonary hypertension (dilatation of pulmonary trunk (▽)). (c) Axial CT contrast-enhanced image shows the interatrial communication (↑).

Fig. 13

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Figure 14. 89 year-old woman with meandering pulmonary vein. CT contrast-enhanced MIP reconstruction shows left upper lobe pulmonary vein with a meandering course.

Fig. 14

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Figure 15. 52 years-old man with Rendu-Osler syndrome. (a) Frontal radiograph of the chest reveals a well-defined lobulated opacity (▲) in the left lower lobe. (b) Contrast-enhanced CT image show a lobulated intensely enhancing lesion (▲) in the left lower lobe. (c and d) MIP images demonstrate a prominent vein (▲) and artery (▲) arising from the lesion.

Fig. 15

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Figure 15. (e and f) Contrast-enhanced CT image show a lobulated intensely enhancing lesion (▲) in the left lower lobe complicated with left hemotherax (▲) and diffuse alveolar hemorrhage contiguous to the lesion (▲). (g and h) Digital subtraction angiographic images before (g) and after amplatz occlusion (h) (▲).

Fig. 16

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Figure 17. 26 years-old patient with unremarkable history, presenting with life-threatening hemoptysis. (a) CT lung window image shows right lower lobe consolidation, surrounding ground-glass images. (b and c) Images of MDCT angiography, (b) axial image showing a vascular anomaly (†) in the right lower lobe adjacent to consolidation, (c) oblique coronal MIP showing an anomalous vessel originating from abdominal aorta (†) going to the right lower lobe, findings suggestive of a sequestration. (d) Volume rendering reconstruction showing the anomalous systemic abdominal vessel (†).

Fig. 17

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Figure 18. 21 years-old man with isolated systemic arterial supply of the lung. (a) CT contrast-enhanced image shows an abnormal vessel (▲) in the lower right lobe. (b, c and d) MIP images demonstrated the abnormal systemic artery and its origin from the abdominal aorta. (e and f) Selective angiography of the anomalous vessel before and after coil embolization.

Fig. 18

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Conclusion

The correct identification of these entities is essential for the accurate diagnosis and appropriate clinical and therapeutic management of the patient.

In patients with hemoptysis without known lung disease, we need to consider the possibility of a congenital malformation (sequestration or isolated systemic arterial supply of the lung).

In studies of pulmonary hypertension we always need to evaluate the presence of anomalous venous drainage, which is very often associated with atrial septal defects.

Personal information

References


