Pictorial Review of Pulmonary Venous Anomalies – What is their Clinical Relevance?

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RELEVANT FINANCIAL DISCLOSURES

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OUTLINE

• Learning Objectives
• Specific Diagnoses
• Conclusion
• References
LEARNING OBJECTIVES

• To illustrate typical imaging features of different pulmonary venous anomalies
• To discuss their clinical implications/relevance
Pulmonary venous anomalies are relatively uncommon, however, may be clinically significant, potentially causing substantial morbidity; therefore it is important to be able to recognize and characterize them.

Imaging is the cornerstone for diagnosis and techniques include chest radiography, echocardiography, computed tomography (CT), and magnetic resonance imaging (MR). CT and MR with IV contrast are the most useful modalities for diagnostic characterization.

Specific Entities discussed in this Educational Exhibit:

- Total anomalous pulmonary venous return (TAPVR)
- Partial anomalous pulmonary venous return (PAPVR)
- Pulmonary arteriovenous malformation (PAVM)
- Pulmonary vein varix
- Variants in pulmonary venous anatomy
- Pulmonary vein stenosis
TAPVR

Four types based on the location of pulmonary venous drainage:

• Supracardiac - most common; pulmonary veins drain most commonly via a vertical vein to the left brachiocephalic vein

• Cardiac

• Infracardiac

• Mixed types
TAPVR

• TAPVR causes massive left to right shunt and usually manifests with neonatal cyanosis

• A patent foramen ovale or ductum arteriosum are necessary for this condition to be compatible with life

• Surgical correction undertaken early in life, and depends on co-existent cardiovascular malformations and whether there is obstruction of the draining vein
PAPVR

• Characterized by at least one pulmonary vein draining into a systemic vein resulting in a left to right shunt

• Most frequently involves anomalous drainage of the right superior pulmonary vein to right atrium or superior vena cava and is frequently associated with a sinus venosus atrial septal defect (ASD)

• In left sided PAPVR, left superior pulmonary vein commonly drains to the left brachiocephalic vein via a vertical vein or to the coronary sinus

• May be clinically silent during childhood, however, pulmonary hypertension is a common complication later in life
PAPVR - RIGHT UPPER LOBE
PAPVR - SCIMITAR SYNDROME

- Rare form of PAPVR which almost always involves the right lung
- Anomalous drainage of a pulmonary vein into the inferior vena cava, portal vein, hepatic veins or the azygos system
- Other associations with this syndrome include: a hypoplastic or aplastic pulmonary artery, ipsilateral pulmonary hypoplasia, cardiac dextroposition and congenital heart diseases
PAPVR - SCIMITAR SYNDROME
PAVM (PULMONARY ARTERIOVENOUS MALFORMATION)

- Abnormal direct communication between a pulmonary artery and vein, bypassing the pulmonary capillary bed
- > 50% of individuals with multiple PAVMs have hereditary hemorrhagic telangiectasia (HHT), whereas 33% of patients with solitary PAVM have HHT
- Most often congenital, classified by number (single or multiple), size and type of arterial supply and venous drainage.
- Most commonly located in the lower lobes
PAVM

- On radiographs, they may resemble a pulmonary nodule.
- On CT, there is simultaneous enhancement of feeding pulmonary artery, nidus and early draining pulmonary vein.
- Require treatment if patients are symptomatic (paradoxic embolization most common presentation, however, numerous PAVMs may present with cyanosis or high output CHF due to right to left shunting) or when the lesion is greater than 2-3 cm.
- Percutaneous transcatheter embolization is the treatment of choice.
PULMONARY VEIN VARIX

• A focal aneurysmal dilation of a pulmonary vein

• Rare entity; can be either congenital or acquired

• Acquired types can develop due to chronic pulmonary hypertension, Behçet’s disease or mitral valvular disease

• Can be classified into three types: saccular, tortuous and confluent

• Complications include rupture, thromboembolism and hemoptyysis, although most small varices are asymptomatic
PULMONARY VEIN VARIX
VARIATIONS OF ANATOMY

• Typically four pulmonary veins and four well differentiated ostia
• Left sided variants basically include convergence of the left pulmonary veins in a common trunk that can be short or long
• Right sided variants less common and mainly include:
  a) one accessory right middle pulmonary vein,
  b) two accessory right middle pulmonary veins,
  c) one accessory right middle pulmonary vein and one accessory right upper pulmonary vein
• Generally inconsequential, however, should be noted prior to cardiovascular surgery or interventional cardiology procedures to decrease the risk of unexpected complications
SEPARATE ACCESSORY RIGHT MIDDLE PULMONARY VEIN
PULMONARY VEIN STENOSIS

- Congenital or acquired
- Congenital pulmonary vein stenosis/atresia can be associated with congenital heart disease or anomalous pulmonary venous drainage
- Acquired stenosis most commonly due to radiofrequency ablation complication (for atrial fibrillation), neoplastic infiltration and fibrosing mediastinitis
- Clinical presentation may include venous congestion, pulmonary edema and alveolar hemorrhage
- Management may include surgical or endovascular correction.
PULMONARY VEIN STENOSIS
CONCLUSION

It is important to recognize and characterize pulmonary venous anomalies, since although relatively uncommon, they may be clinically significant, potentially causing substantial morbidity.

Accurate diagnosis is crucial for optimal management.
REFERENCES


THANK YOU FOR YOUR ATTENTION!

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