Vascular malformation and variants: Circulation through crooked paths

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Learning objectives

To characterize common and rare vascular anatomical variants and malformations, including potential pitfalls and differential diagnoses.
Background

Vascular anomalies result from embryological developmental deformities and, although they are present since birth, they are often asymptomatic and discovered incidentally in adulthood. Knowledge of normal anatomy and recognition of these vascular variants is crucial to correct diagnosis; in fact, these anomalies are frequent pitfalls in imaging interpretation.
Findings and procedure details

We collected several patients with vascular malformations (previously known or incidental) to review its imaging findings on x-ray and computed tomography (CT).

Our cases comprise certain relatively common vascular variants, such as aberrant right subclavian artery and left renal vein variants, but also some rarer anomalies, such as interruption of inferior vena cava (IVC) and right aortic arch.

Although these variants are frequently asymptomatic, they may be important pitfalls: for example, with interruption of IVC, a prominent azygous or hemiazygous vein may simulate a mediastinal mass or adenopathy. Accurate characterization requires a thorough analysis of contiguous CT sections, usually facilitated by intravenous contrast agent administration and coronal and sagittal reconstructions.

On the other hand, other variants may become symptomatic during certain procedures, and complicate its execution: jugular vein agenesis and left sided superior vena cava (SVC), for example, can cause distress while central venous line or pacemaker placement. Left renal vein variants may also cause significant problems in retroperitoneal surgery or interventional procedures.

Internal jugular vein agenesis (fig. 1-2)

Internal jugular vein is a main contributor to venous drainage from intracranial structures. It runs downwards through the neck in the carotid sheath in close approximation with the internal carotid artery, from the sigmoid sinus until it joins the subclavian vein to form the brachiocephalic vein. Unnoticed agenesis of internal jugular vein can cause significant complications when placement of a central venous line, as it is the most common vein used in central venous access. Also during neck procedures, such as dissection of metastatic lymph nodes in head and neck cancers, it is crucial to recognize this variant in order to avoid interruption of the alternative pathways (external jugular vein) and to prevent injury to the contralateral side.

Aberrant right subclavian artery (fig 3-5)

Aberrant right subclavian artery, also known as arteria lusoria, is the most common arch anomaly, occurring in about 1%-2% of the population. It arises as the last branch of the aortic arch, rather than from the brachiocephalic artery, and crosses the mediastinum obliquely from left to right, passing most commonly behind the esophagus and trachea. In these patients, the brachiocephalic artery has a smaller diameter than usual, as it corresponds actually only to the right common carotid artery. This variant is usually
asymptomatic and found incidentally. However, its origin is frequently dilated, known as the diverticulum of Kommerell, and can compress and displace the adjacent esophagus and cause dysphagia (*dysphagia lusoria*). Its recognition is important as it may be misdiagnosed as a mediastinal mass or an aneurism of the aortic arch.

**Right aortic arch** (fig 6-9)

Right aortic arch occurs in approximately 0.05%-0.1% of population and consists of an aortic arch that passes to the right of the trachea and esophagus. It may be associated with mirror image branching, which is frequently associated with congenital heart disease (such as tetralogy of Fallot), or with aberrant left subclavian artery, which is similar to a normal left arch with an aberrant right subclavian artery.

**Persistent left superior vena cava** (fig 10-14)

Persistent left superior vena cava is found in 0.3% of general population and in 5% of patients with congenital heart disease. In majority of cases, the right SVC is also present (SVC duplication). A persistent left superior vena cava may be misdiagnosed as an adenopathy and it can cause problems to a pacemaker or line placement. Usually it can be seen the left superior intercostal vein in the left mediastinum, from the hemiazygous vein to the abnormal left superior vena cava.

**Azygous vein arch and azygous lobe** (fig 15-19)

Azygous fissure (azygous vein surrounded by a double fold of visceral and parietal pleura) results from incomplete migration of the right posterior cardinal vein. The resulting azygous vein, laterally displaced, forms the azygous lobe, which is present in about 1%-2% of population. It has no clinical significance and it is usually detected by radiologists in routine chest radiographs. The term "lobe" is not enterily correct, as it does not have its own bronchus and there is no alteration in segmental anatomy of the lung (it is usually supplied by branches of the apical segmental bronchus) Its diagnose on CT is not difficult when the azygous vein has a transverse course and is intercepted in one slice, but when its path is more vertically can be misdiagnose as an adenopathy (specially in CT studies without IV contrast). In fact, IV contrast and close evaluation through consecutive slices allows the correct diagnosis. Azygous vein usually ends in SVC or, rarely, in right brachiocephalic vein. Although asymptomatic, its appearance on radiographs and CT may simulate disease, so it is crucial its detection and differentiation from pathologic entities.

**Inferior vena cava anomalies** (fig 20-24)

IVC anomalies comprise numerous anatomic variants, as its embriology requires a complex process of anastomosis between three paired embryonic venous systems.
(posterior cardinal, subcarinal and supracardinal). IVC congenital anomalies can be classified as pre-renal, as interruption of IVC with azygous continuation (prevalence 0.6%-1.5%); and post-renal, as double IVC (prevalence 0.2%-3%) and left-sided IVC (prevalence 0.2%-0.5%). Additionally, and because the embryologic process is so complex, more than one anomaly can coexist, resulting in a great number of combinations, which are even rarer than the simple variants themselves. Our case is of a 69-year old women, asymptomatic, who performed a chest CT and IVC interruption with hemiazygous continuation was incidentally found. The prevalence of this combination is not established in literature, being more common the presence of a double IVC with hemiazygous continuation (usually associated with retroaortic right or left renal vein). The hemiazygous vein crosses posterior to the aorta at the level of T8-T9 vertebral bodies to join the azygous vein (or alternatively joining a persistent left SVC or continuing as accessory hemiazygous vein to the left brachiocephalic vein) Besides its implication in interventional and surgical procedures, it is important to avoid diagnostic pitfalls, as aberrant vessels can simulate mediastinal masses or retrocrural and retroperitoneal adenopathies. IVC variants are frequently associated with other abnormalities, such as congenital heart disease, asplenia or polysplenia syndromes.

**Left renal vein variants** (fig 25-31)

Left renal vein variants include retroaortic left renal vein (prevalence 2%) and circumaortic venous ring (prevalence 10%) and result of persistence of the dorsal limb of the embryonic left renal vein and of the dorsal arch of the renal collar. If unrecognized, these anomalies may cause significant problems in retroperitoneal surgery or interventional procedures. On the other hand, compression of a retroaortic renal vein can cause urologic symptoms due to increased pressure, such as hematuria and inguinal or flank pain. Retroaortic left renal vein has also been associated with varicocele, as its compression (similar to a "nutcracker phenomenon", in which a normal left rein vein is compressed between the aorta and the superior mesenteric artery) can impair venous drainage and enlarge and revert blood flow in the gonadal veins.
Fig. 1: Right internal jugular vein agenesis: note the absence of internal jugular vein on the right and the normal aspect of internal jugular vein on the left, in intimate correlation with the internal carotid artery.

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Fig. 2: Right internal jugular vein agenesis: note the absence of internal jugular vein on the right and the normal aspect of internal jugular vein on the left, in intimate correlation with the internal carotid artery.

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**Fig. 3:** Aberrant right subclavian artery, or arteria lusoria, arising from the aortic arch.

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Fig. 4: Aberrant right subclavian artery and its origin in the aortic arch.

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Fig. 5: Coronal reconstructions showing aberrant right subclavian artery, its origin in the aortic arch and its course upwards and to the right.

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Fig. 6: Right aortic arch

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Fig. 7: Right aortic arch: its origin in the left ventricle and its proximal segments.

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Fig. 8: Right aortic arch and the origin of a left brachiocephalic artery, which will give origin to left common carotid artery and left subclavian artery.

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Fig. 9: Right aortic arch and its branches: R SCA (right subclavian artery), L SCA (left subclavian artery) and L CCA (left common carotid artery).

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Fig. 10: Left superior vena cava: coronal reconstructions

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Fig. 11: Left superior vena cava (arrow) with a pacemaker line.

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Fig. 12: Left superior vena cava (*) found incidentally while pacemaker introduction. Arrows show the course of the pacemaker line.

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Fig. 13: Left superior vena cava: coronal reconstructions

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**Fig. 14:** Left superior vena cava (*). Arrows show left superior intercostal vein draining into the left-sided superior vena cava.

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**Fig. 15:** Azygous vein laterally displaced, forming the azygous lobe.

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Fig. 16: Azygous vein and fissure. When the azygous vein course is not transverse, it can simulate a nodule.

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**Fig. 17:** Azygous vein laterally displaced, forming the azygous lobe.

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Fig. 18: Azygous fissure on chest radiograph

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Fig. 19: Azygous vein and fissure in coronal reconstructions

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**Fig. 20:** Inferior vena cava interruption with hemiazygous continuation. The abnormally dilated hemiazygous vein, located on the left side of the aorta, can simulate an adenopathy; IV contrast injection and the absence of a normal inferior vena cava help with the correct diagnosis.

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**Fig. 21:** Inferior vena cava interruption with hemiazygous continuation. The abnormally dilated azygous vein, located on the right side of the aorta, can simulate an adenopathy; IV contrast injection and following its course through multiple contiguous slices help with the correct diagnosis.

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Fig. 22: Inferior vena cava interruption with hemiazygous continuation. Hemiazygous joining the azygous vein (red arrow) can simulate an adenopathy. Green arrow shows the normal supra-hepatic segment of the inferior vena cava.

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**Fig. 23:** Inferior vena cava interruption with hemiazygous continuation. Arrow shows the azygous arch.

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**Fig. 24:** Inferior vena cava interruption with hemiazygous continuation. Renal veins joining the hemiazygous vein (retroaortic right renal vein)

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Fig. 25: Retroaortic left renal vein

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Fig. 26: Retroaortic left renal vein: note its compression behind the aorta.

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Fig. 27: Retroaortic left renal vein

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Fig. 28: Retroaortic left renal vein

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**Fig. 29:** Retroaortic left renal vein in coronal reconstructions

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**Fig. 30:** Circumaortic left renal vein: left renal vein passing normally anterior to the aorta and a supranumerary left renal vein passing behind the aorta.

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Fig. 31: Circumaortic left renal vein: left renal vein passing normally anterior to the aorta and a supranumerary left renal vein passing behind the aorta.

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Conclusion

Vascular anomalies are important imaging pitfalls and need to be correctly interpreted and recognized.
References


