Superior vena cava obstruction: causes and imaging findings.

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Learning objectives

- To review the causes of superior vena cava syndrome (SVCS).

- To study the relationship between hemodynamic changes and radiological findings.
Background

Superior vena cava syndrome is the result of compression, invasion, or thrombosis of this thin-walled blood vessel, which leads to impaired venous drainage and congestion of the head, neck and upper extremities [1, 2]. Clinical manifestations include facial and neck swelling and distended neck veins. Other symptoms, such as dyspnea, dysphagia, cognitive dysfunction, and severe headache are often associated with this disorder.

Correlation of imaging studies with clinical findings suggests that the severity of the symptoms depends on the level of obstruction (above or below the level of the azygos arch) and the development of rich collateral network. Contrast-enhanced computed tomography (CT) is one of the most useful imaging modalities in the evaluation of SVCS. CT can show the level of the obstruction even in patients relatively asymptomatic [3].

Different causes for this syndrome, specific physiopathology and collateral circulation have been analyzed in this review.

Radiological findings on CT and phlebography (before and after treatment) have also been reviewed.

Conventional venography, the historical reference standard, is a more invasive procedure and should only be performed whenever invasive therapeutic procedures are planned. It has been replaced by noninvasive imaging techniques including CT.

MDCT is widely considered to be a reliable technique for defining the etiology, grade of severity, and planning invasive treatment.

Regardless of its cause, the CT diagnosis of superior vena cava obstruction includes several radiological criteria [4]:

- Absent or diminished opacification of the SVC.
- Severe narrowing of the SVC.
- Collateral routes of venous drainage.
- Intraluminal filling defect, if thrombosis is associated.

CAUSES OF SVC OBSTRUCTION
This syndrome is usually an acquired condition and the different causes are:

**Malignant:** it is the main cause. It may be primary neoplasm (fig.1-6) or mediastinal nodal metastases (fig. 7). Cancer is the most common underlying cause of superior vena cava obstruction. Lung cancer, either non-small or small cell cancer, can directly invade superior vena cava (fig. 1). Lymphoma, either Hodgkin disease or non-Hodgkin lymphoma, tends to compress the vessel. Other mediastinal tumors, including malignant thymoma, germ cell tumors, and metastases or pleural cancer, such as mesothelioma, can also result into superior vena cava syndrome [1].

Most primary malignant tumors of the vena cava, such as leiomyosarcoma or angiosarcoma, usually affect inferior vena cava. Involvement of the superior vena cava is extremely uncommon.

The presence of arterial enhancement of the thrombus is highly suggestive of tumoral thrombus.

**Benign:** this category includes infection, fibrosing mediastinitis (fig.8-11), Behçet disease, retrosternal goiter, aortic aneurysm, benign tumors, mediastinic hematoma (fig. 12 - 13), sarcoidosis or radiation-induced fibrosis [5].

Fibrosing mediastinitis is a rare histologically benign disorder caused by proliferation of collagen tissue and fibrosis in the mediastinum. It may be idiopathic, caused by an abnormal immunologic response to *Histoplasma capsulatum* infection or tuberculosis, or it may be related to retroperitoneal fibrosis, particularly in its diffuse form.

On CT, fibrosing mediastinitis appears as an ill-defined soft tissue mass obliterating the normal fat. The middle mediastinal compartment is most commonly affected. A more focal form with calcified mass has also been described (fig.8) [6].

Behçet disease is a rare systemic disease that most commonly affects patients of Mediterranean, Middle Eastern, and Asian ancestry and causes recurrent genital and oral ulcers and it might cause uveitis too. It is an inflammatory disorder associated with leukocytoclastic vasculitis affecting the arteries and veins of the systemic and pulmonary circulations. Superior vena cava stenosis or occlusion may result from thrombophlebitis or fibrosing mediastinitis, although involvement of major veins is uncommon.

**Iatrogenic:** it is the most common etiology after malignant causes. The increasing of intravascular devices has extended the incidence of SVC syndrome arising from benign etiologies [1]. Neointimal hyperplasia secondary to repeated trauma of the tip of catheter
associated with the stasis induced by its presence, are the main predisposing factors. Large central venous lines, such as Hickman catheters, dialysis catheters or parenteral nutrition catheters, and transvenous pacemaker wire (fig.14), have been associated with SVCS.

**COLLATERAL CIRCULATION**

In case of chronic occlusion of SVC collateral pathways must develop to maintain venous drainage to the heart [1]. The development of collateral vessels protects the patient to develop more severe symptoms.

Most veins draining the upper half of the body (brachycephalic veins, superior vena cava, and azygos-hemiazygos system) are connected by a network of smaller venous plexuses that are normally collapsed.

The presence of collateral veins is an accurate predictor of SVC syndrome, with a sensitivity of 96% and a specificity of 92% [7]. The extent and location of collateral veins are extremely variable and often involve the chest, abdomen, and pelvis. The pattern of collateral pathways can be predicted depending on the level of obstruction.

Multi-detector row computed tomography (MDCT), with its multiplanar and volume rendered images can be used as an interpretative aid to depict the different collateral pathways from the site of the obstruction. Although axial images allow evaluation of potential causes of superior vena cava obstruction, multiplanar reconstruction provides invaluable information about the level and degree of the obstruction, the length of the affected segment, and the presence or absence of intraluminal clot distal to the obstruction, thereby allowing the interventional radiologist to choose the optimal treatment option [4].

**Thoracic collaterals:**

On MDCT, several major plexuses in the thorax are enlarged and become visible. Although many factors influence in the opacification of venous collaterals, depiction of dominant collateral channels may provide a clue to the level of venous occlusion and the direction of venous flow [8]. The most commonly visible venous collaterals are [1, 9]:

- Azygos-hemiazygos-hemiazygos accessory system (fig.2): it serves as a conduit for venous blood from intercostal and lumbar veins to SVC and is the main collateral
pathway. The azygos drains via the azygos arch at the level of the right main bronchus (fig 7, 9 and 11) [2]. If the level of superior vena cava obstruction is above the azygos arch, anterograde flow from the azygos to the right atrium is seen, with abrupt transition between a densely opacified azygos superiorly (caused by drainage of intercostal veins) and unopacified azygos inferiorly. If the obstruction is below the arch, retrograde flow from the azygos-hemiazygos system to IVC is seen, with opacification through the entire length of the azygos-hemiazygos system (fig.5) [8].

- **Vertebral and subscapular plexuses:** the vertebral venous plexus of paravertebral, intervertebral, and epidural veins provide collateral pathways to the azygos system or to the IVC through the ascending lumbar veins (fig. 3 and 9).

- **Mediastinal, esophageal and diaphragmatic venous plexuses:** they include mediastinal, pericardial, and pericardiophrenic veins (fig.9); and they drain to inferior vena cava either inferiorly through inferior phrenic veins or via transhepatic collaterals.

- **Lateral thoracic and superficial thoracoabdominal venous plexuses:** the internal and lateral thoracic veins communicate with thoracoepigastric and superficial epigastric veins. Chest wall and breast collaterals can also brightly opacify as a sign of superior vena cava obstruction (fig. 5, 7, 9, 11 and 14) [1].

**Abdominal venous collaterals:**

In addition to enhancing round or tortuous vascular channels in the abdominal wall (fig.9), perfusion abnormalities in the liver and the so-called "hot spot" (intense opacification in the segment IV on CT) can also be seen; both as a result of the communication between superficial epigastric veins and left portal vein (fig.15).

Another possible collateral channel involves the inferior phrenic veins and subcapsular hepatic veins with intense opacification of the subdiaphragmatic portion of the liver.

**Unusual collateral pathways:**

There are other less common pathways, such as systemic to pulmonary shunt which are usually the result of SVC obstruction from malignant causes. The pathway consists of mediastinal connections between the innominate veins and the superior pulmonary veins via the bronchial venous plexuses around the airways, hilar vessels, and pleura [4].
To enable a good interpretation of the radiological findings before and after the treatment, it is therefore essential to understand the hemodynamic and collateral pathways in SVCS.

**TREATMENT**

Endovascular treatment by stenting or PTA of SVCS, is a simple and safe procedure to restore the patency of superior vena cava. It should be indicated in most cases as first-line treatment because it does not interfere with other treatments and provides an urgently needed relief of symptoms [11-12].

Endovascular stent is an excellent initial treatment for SVCS of malignant cause, mainly for the immediate relief of symptoms (fig. 2, 4 and 6) and because these patients have a short life expectancy.

Endovascular prostheses are very effective in the treatment of benign causes of SVCS (fig.10), including patients with pacemakers [1], but stent implantation requires prophylactic anticoagulation or antiplatelet aggregation for long periods of time in order to avoid thrombosis as a complication (fig.11). Therefore, angioplasty treatment is chosen as the first option in benign causes, delaying the stent treatment as long as it is possible.

MDCT allows (by seeing or not opacification of the lumen) to assess if the stent is occluded (fig.3 and 11). Furthermore, collateral circulation is an indirect sign of a nonfunctioning stent.
Imaging findings OR Procedure details

Different cases of superior vena cava syndrome with distinct etiology at our institution were reviewed. Radiological findings on 16-row MDCT were related to phlebographic findings before and after treatment.
Conclusion

- SVCS is due to multiple causes and is, most commonly, an acquired condition.

- Haemodynamic changes in SVCS explain the different collateral circulation pathways and radiological findings. Those collateral pathways can be predicted depending on the level of obstruction.

- MDCT is a useful tool for diagnosis and treatment planification of SVCS. Phlebography is used as prelude to endoluminal treatment.
References


