

Superior vena cava syndrome from a tomographic image perspective. Case report

Síndrome de vena cava superior desde una perspectiva de la imagen tomográfica. Presentación de caso

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Summary

The superior vena cava syndrome is a set of signs and symptoms resulting from the compression or occlusion of the superior vena cava, most commonly secondary to external compression by a neoplastic process. Imaging plays a fundamental role in the diagnosis and identification of the syndrome. We present the case and images of a patient with superior vena cava syndrome secondary to mediastinal adenopathy, related to possible metastasis from uroepithelial carcinoma of the bladder.

Resumen

El síndrome de la vena cava superior es un conjunto de signos y síntomas que resultan de la compresión u oclusión de la vena cava superior, más comúnmente secundario a procesos neoplásicos. Las imágenes juegan un papel importante en el diagnóstico e identificación de este síndrome. Se presentan el caso y las imágenes de un paciente con síndrome de vena cava superior secundario a adenopatía mediastinal, relacionado con posible metástasis de carcinoma uroepitelial de vejiga.

Introduction

Superior vena cava syndrome (SVCS) is a set of signs and symptoms resulting from compression or occlusion of the superior vena cava (SVC). This is a pathology associated with significant morbidity and mortality (1).

SVCS is secondary to impaired venous drainage of the head, neck, upper extremities, and thorax due to SVC obstruction.

SVC obstruction most often results from extrinsic compression caused by a neoplastic process, such as a tumor arising on the right side of the main or upper lobar bronchus or from large mediastinal lymphadenopathy (paratracheal, subcarinal, and/or hilar), and less commonly from intravascular neoplastic invasion or thrombus formation.

Blood flow is redirected through multiple collateral blood vessels. Vessels bypass the obstruction through the azygos vein or inferior vena cava depending on the degree of obstruction. Over time, the caliber of these collateral vessels increases, leading to increased blood flow (2).

Imaging plays a key role in confirming the diagnosis of SVCS and identifying its underlying cause. Non-invasive imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) are used. CT is most commonly used, as it is the most widely available modality with a short acquisition time that reduces respiratory and cardiac motion artifacts. In addition, multidetector CT with contrast medium and multiplanar reconstructions in association with 3D volume rendering are useful in identifying the site, extent of occlusion, intravascular thrombus and collateral circulation, in addition to the root cause, and is therefore considered a superior imaging modality to conventional venography (3).

Case report

An 81-year-old patient with a history of high-grade uroepithelial carcinoma of the bladder undergoing surgical and chemotherapeutic management, presents with progressive edema of the face, neck and upper extremities, associated with telangiectasias in the anterior thorax, of three months of evolution. A CT with intravenous contrast medium was performed with arterial and venous phases, with multiplanar and three-dimensional reconstructions (Figure 1), showing mediastinal adenopathy at 4R level, with external compression of the SVC, associated with thrombosis of the same extending to the right brachiocephalic and innominate veins, with multiple collaterals through intercostal veins (anterior, posterior and superior), internal thoracic vein and lateral vein on the left side; In addition,

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Figure 1. CT image sequence: a) axial, b) sagittal, c) coronal and d) three-dimensional reconstruction. Mediastinal adenopathy is visualized at 4R level (red arrowhead) that generates compression on the SVC (red arrow), as well as filling defect extending through the right brachiocephalic and innominate vein in relation to thrombosis (blue arrow), with presence of abundant intercostal and thoracic collaterals on the left side (yellow arrow).

bilateral pleural effusion predominantly right with right adjacent passive atelectasis. Management with anticoagulation, diuretics and palliatives is considered in committee.

Discussion

SVCS comprises a broad clinical spectrum ranging from asymptomatic cases to life-threatening emergencies.

In the past, SVCS was preferentially caused by compression of the SVC by syphilitic aortic aneurysms and mediastinal lymphadenopathy due to tuberculosis. Today, compression or invasion of the SVC by malignant tumors (about 60 %) and thrombosis or stenosis caused by central lines or medical devices (about 30-40 %) predominate. The most frequent tumors are lung cancers, which account for 70 % of all SVCS cases, especially microcytic cancers and lymphomas. Other underlying pathologies, such as fibrosing mediastinitis, are rare (4, 5).

The method of choice for the diagnosis of SVCS in emergency and elective conditions is CT with contrast medium with multi-phase imaging, since it has a high sensitivity and specificity for the diagnosis of this entity and also allows to reliably define the underlying cause and additional findings, such as intrapulmonary metastases, pleural effusion and lymphadenopathy, as well as being very useful in the evaluation of collateral circulation and for the planning of interventional procedures (5).

Regardless of its cause, CT diagnosis of SVC obstruction includes lack of SVC opacification, an intraluminal filling defect or severe narrowing of the SVC, and visualization of collateral vascular channels; therefore, they can be classified into direct signs: absence of opacification of the central veins (innominate or SVC), either due to extrinsic compression or an intraluminal repletion defect; the increase in their caliber, due to the presence of intraluminal thrombus or clot and stenosis of the vessel lumen secondary to fibrosis, usually due to the use of endovenous catheters. Indirect signs can also be visualized, such as opacification of the collateral venous plexuses, increased in

caliber and with a tortuous course, which under normal conditions should be collapsed. This sign implies a gradual establishment of obstruction, which allows the collateral vascular network to accommodate the flow coming from the SVC (6). The presence of collateral vessels is highly suggestive of SVCS, with a sensitivity of 96% and a specificity of 92% (7).

If the site of SVC obstruction is above the azygos arch, antegrade flow from the azygos into the right atrium is observed, with an abrupt transition between a densely opacified azygos above the arch and a non-opacified inferior azygos vein. If the obstruction is below the arch, all the azygos and hemi-azygos veins become intensely opacified as blood flows retrograde into the inferior vena cava. The vertebral venous plexus of paravertebral, intervertebral, and epidural veins provides collateral pathways to the azygos system or to the inferior vena cava via the ascending lumbar veins. Other collaterals that can be visualized in the SVCS are the mediastinal, pericardial and pericardiophrenic veins that drain into the inferior vena cava, either inferiorly through the inferior phrenic veins or through transhepatic collaterals. The internal thoracic veins and lateral thoracic veins communicate with the thoracoepigastric and superficial epigastric veins. Chest wall and breast collateral veins may also become intensely opacified as a sign of superior vena cava obstruction.

The radiologist should be familiar with abdominal CT findings suggestive of SVC obstruction. In addition to enhancing round or tortuous vascular channels in the abdominal wall, perfusion abnormalities in the liver result from communication between the superficial epigastric veins and the left portal vein. On CT there is intense opacification of the anterior square lobe. The area of enhancement is characteristic in both position and shape and should not be confused with a hypervascular mass (6).

The pressure in the SVCS is -4.5 to -6 mm Hg, the intrathoracic and neck veins are open during inspiration, which facilitates venous return, and are collapsed during expiration. In SVCS the venous pressure increases in the range of 20 to 40 mm Hg, which causes an increase in hydrostatic pressure. The intensity of symptoms depends on the degree of compression and the speed of onset of SVCS.

At the onset of the clinical picture, acute obstruction of SVC flow transiently decreases cardiac output. Within hours, venous pressure increases via collateral veins to the azygos, paravertebral, internal mammary and intercostal veins, to name a few, which dilate, a mechanism that takes several weeks. This compensatory mechanism may be limited if the compression of the SVC is below the azygos vein, worsening the clinical manifestations associated with this syndrome (8).

Pressure measurements may also be useful in determining the severity of superior vena cava obstruction (7).

Contrast-enhanced MRI could be used as an alternative diagnostic tool in case of intolerance to iodine contrast media.

Previously, cavography and phlebography of the upper extremities were considered the diagnostic gold standard. Its strength was the detection of intraluminal processes such as thrombus formation and determination of the degree of obstruction, as well as assessment of collateral pathways, but compared with contrast CT, it is less useful in detecting causes of obstruction other than thrombus. With modern contrast-enhanced CT or MRI techniques, cavography/phlebography is restricted to interventional therapy. If cavography is performed, at least two projections (posteroanterior and lateral) are necessary. Findings may appear normal in one projection, but be pathologic in an additional projection.

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