CT Findings in Superior Vena Cava Obstruction

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The clinical manifestations of SVCS were first described by Hunter in 1757, secondary to a syphilitic aneurysm of the ascending aorta. SVCS usually has an insidious presentation and eventually leads to a collection of signs and symptoms that together are diagnostic of the disorder. Obstruction of the SVC may result from external compression or intrinsic thrombotic disorders, and leads to proximal venous hypertension of varying magnitude. Clinically, the syndrome is manifested by: (1) the progressive dilatation of the veins of the upper trunk, arms, neck, and head; (2) increasing edema and plethora of the face, neck, upper torso, and breasts (Fig 1); (3) CNS disturbances with visual symptoms, syncope, dizziness, headaches, altered state of consciousness, and somnolence; (4) cyanosis and conjunctival edema; and (5) edema of the trachea and esophagus with respiratory distress and dysphagia.

The development of the SVCS relates to the nature of the blood vessel as well as to the surrounding mediastinal anatomy. The SVC is a thin-walled structure without valves that drains the returning blood flow of the head and upper half of the trunk. It measures approximately 7 cm in length and has a varied cross-sectional shape. The greatest diameter of the normal SVC is generally about 1.5 cm, but can be as much as 2.0 cm.

The intravascular pressure of the SVC is low, a reflection of the normally low systemic venous pressure. The SVC is firmly bound in the right anterosuperior mediastinum and is in contact with the ascending aorta, right pulmonary artery, left atrium, right pulmonary veins, and the right main stem bronchus. It is flanked by a network of lymph nodes and small lymphatic vessels. Lymph drainage from the right thoracic cavity and the lower part of the left cavity passes through these nodes.

The azygos vein is the major tributary of the SVC and is formed by the lumbar veins at the level of the diaphragmatic crura, and ascends to enter the dorsal aspect of the mid-SVC. Small pericardial and mediastinal veins may drain into the intrapericardial portion of the SVC.

The lower half of the SVC is covered by the parietal pericardium, and its wall is partially lined by the visceral pericardium. There is relative fixation of the SVC due to its intrapericardial location, leaving little room for its displacement by a space-occupying lesion. The vessel is, however, susceptible to pericardial or cardiac abnormalities.

Compression of this low-pressure, thin-walled vessel leads to compromise of its lumen (Figs 2 and 3). When the lumen is completely obliterated, the signs and symptoms of SVCS become evident, especially if the azygos vein orifice is also obstructed. However, partial obstruction occurring slowly may allow enough collateral circulation to develop so that the obstruction may not be clinically apparent.

In the event of SVC obstruction, there are four main routes of collateral circulation between the SVC and IVC: azygos/hemiazygos, internal mammary, vertebral, and thoracoepigastric/lateral thoracic veins. Superior intercostal and paraesophageal veins may also serve as part of the collateral circulation. Blood may be diverted to the peri umbilical veins and subsequently to the

ABBREVIATIONS

CNS, central nervous system
IVC, inferior vena cava
LIV, left innominate vein
MRI, magnetic resonance imaging
RUL, right upper lobe
SVC, superior vena cava
SVCS, superior vena cava syndrome
SUPERIOR VENA CAVA OBSTRUCTION

Fig 1. Massive edema of the chest wall in a patient with SVC obstruction. Pleural effusions (E) are present.

left portal vein along the ligamentum teres by superficial thoracoabdominal and epigastric veins, accounting for the "hot spot" projected over the liver on radionuclide scans in such cases. In cases of SVC obstruction, Ishikawa et al. described an abnormal area of enhancement in the liver on contrast-enhanced CT, representing collateral flow.

ETIOLOGY

The overwhelming majority of cases of SVC obstruction are due to malignant disease. Iatrogenic manipulation has recently become increasingly frequent. The pulmonary artery catheter, peritoneovenous shunt, hyperalimentation line,

Fig 2. Mediastinal lymphadenopathy causing narrowing of the SVC in a patient with lymphoma.

Fig 3. Displacement, distortion, and compression of the LIV, SVC, trachea (t), and aorta (ao) by massive mediastinal lymphadenopathy (Tu) following right pneumonectomy for a recurrent bronchogenic carcinoma.
central venous pressure monitor, and transvenous pacemaker have all been associated with SVCS. The wall of the SVC may be compressed or directly invaded by tumor. Tumor thrombus may occlude the vessel (Fig 4) or the lumen may be narrowed and encased by nodal metastases.

McIntire and Sykes, in a review of cases of SVCS before 1949, showed that only one third were caused by primary intrathoracic malignancy. One third were secondary to aortic aneurysm (now a rare cause). Compression by scar tissue (chronic mediastinitis) accounted for 15% and metastatic disease for 3%.

Recent reviews have shown the percentage of cases of SVCS from malignant tumor has been steadily increasing. Today SVCS has become virtually synonymous with malignant obstruction. Reviews indicate a primary malignant etiology for SVCS in 65% to 97% of the patients. Lochridge et al reported that 82% of the patients with malignant etiology had bronchogenic carcinoma, 12% lymphoma, and 6% metastatic disease. Ooizumi et al reported an incidence of SVCS of 4% among all patients with lung cancer and 8% of patients with small cell type. Shimm et al stated that 5% to 15% of lung cancers, 5% of lymphomas, and 1% of pulmonary metastases result in SVCS. The most common metastatic
neoplasm to cause SVCS has been reported to be breast carcinoma.\textsuperscript{11}

SVC obstruction is uncommon in childhood and adolescence, and is usually iatrogenic. The predominant primary cause of SVCS in these age groups are mediastinal tumors, the most common offender being the lymphoproliferative neoplasms.\textsuperscript{16}

Of the minority of cases of SVCS with a benign etiology, about 75\% are secondary to mediastinal granulomatous disease, and the remainder are caused by such disorders as retrosternal goiter and aortic aneurysms (Fig 5).\textsuperscript{17}

**DIAGNOSIS**

In recent years there has been controversy over the diagnosis and management of SVC obstruction. Selection of an appropriate diagnostic procedure should be based on whether the following critical information is required: (1) definition of the site and extent of obstruction; (2) routes of collateral circulation; and (3) acquisition of tissue for a histopathologic diagnosis.

Plain chest radiographs are of limited diagnostic value. Mediastinal widening is reported to be the most common abnormality on the chest radiograph in SVC obstruction.\textsuperscript{10} Other abnormalities that may be seen include pleural effusion, right hilar mass, bilateral diffuse infiltrates, cardiomegaly, RUL collapse, and anterior mediastinal mass. SVC obstruction may also result in rib notching. It is important to remember that a normal chest radiograph is not inconsistent with the diagnosis of SVC obstruction.\textsuperscript{10,18}

Esophagography may demonstrate filling defects compatible with esophageal varices. Classically, the varices are “downhill,” representing collateral circulation through enlarged upper esophageal veins, bypassing the obstructed SVC to reach the IVC.

SVC phlebography has been the standard procedure for the diagnosis of SVC obstruction and provides useful information. The route and extent of collateral circulation is well demonstrated, and may be of prognostic importance in predicting clinical response. Opacification of the innominate veins by phlebography is useful for planning a venous bypass and for designing radiation portals. Collateral vessels are clearly demonstrated with contrast-enhanced CT, and can better demonstrate the location and extent of both tumor and thrombus, thus offering more information to the radiotherapist.

A recent review has found SVC venography to be of limited value compared to CT.\textsuperscript{10} The venogram is seldom needed since it offers no information about histology and the diagnosis is often clinically apparent. Furthermore, elevated venous pressure in the arms may lead to difficulty...
in controlling bleeding at the puncture site. Because of the ancillary benefits of the CT scan, a previously performed venogram should not obviate the need for CT.

Diagnosis of SVC obstruction may also be suggested by scintigraphy, which also demonstrates obstruction of flow and collateral vessels. However, the resolution of scintigraphy is lower than venography, and it also fails to demonstrate the pathologic anatomy of the obstruction.

The diagnosis of SVC obstruction by contrast-enhanced CT requires that at least two criteria be met. First, there must be diminished or absent
opacification of central venous structures such as the innominate vein or SVC inferior to the site of obstruction. Second, collateral venous routes must be opacified. The fulfillment of either criteria alone is not sufficient for an accurate CT diagnosis of venous obstruction. It is well known that dilution of the contrast medium by unopacified blood and its displacement by laminar flow may simulate an intraluminal filling defect. The absence of opacification of collaterals helps to make the distinction. Conversely, one may see opacification of collateral veins in the absence of mediastinal obstruction, ie, with normal opacification of the SVC and innominate veins. Thus

Fig 8. Bronchogenic carcinoma. (A) Narrowing and encasement of both the superior vena cava (SVC) and right main bronchus (B) by a large right upper lobe mass with mediastinal extension. (B) CT digital phlebogram confirms the presence of a tapered narrowing of the encased SVC (arrows). Ao, aorta; C, collateral veins. (Reproduced with permission from Moncada R et al and the American Roentgen Ray Society.)
the diagnosis of SVC obstruction requires the presence of intraluminal filling defects (Fig 6), or extrinsic compression or encasement of major thoracic venous channels, combined with collateral vessel opacification (Fig 7).

The diagnosis of SVCS can be reliably made by chest CT, which provides detailed anatomic information of the entire mediastinum and is less invasive than other methods, especially bronchoscopy and mediastinoscopy. It may provide the information needed to plan a safe route for percutaneous fine needle biopsy and to direct bronchoscopy toward the area of interest for specimen retrieval. CT is useful in planning radiation portals and is the procedure of choice for follow-up after treatment. It may also be employed to evaluate the patency of a venous bypass.

The recent addition of MRI to the diagnostic armamentarium provides a potential alternative modality in selected cases. The ability of MRI to demonstrate vascular structures without the use of intravenous contrast medium makes it an attractive alternative. However, unstable patients may not be able to cooperate with this somewhat lengthy examination. Recent advances in cardiac and respiratory gating as well as continued advances in fast scanning may make this the procedure of choice for the future, but at present more experience is needed before adopting this modality.

CT digital phlebography has been reported as a valuable adjunct to axial CT scanning in SVC obstruction (Fig 8). It provides an AP bidimensional image of the thoracic veins as well as transverse axial images. Venography can usually be avoided when this combination technique is employed. The CT digital phlebogram can also reveal patency of a venous bypass of a SVC obstruction. We believe that the combination of CT digital phlebography and axial CT scanning is currently the most efficient, informative, and cost-effective technique available for the diagnosis and management of SVC obstruction.

REFERENCES