

Compensatory dilatation of the Azygos Venous system Secondary To Superior Vena Cava Occlusion

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ABSTRACT

Superior vena cava (SVC) occlusion can be clinically recognized in the acute setting when the stenosing process does not allow the development of collateral venous channels, which guarantee the venous drainage to the right heart. On the contrary, when the obstruction develops progressively, the diagnosis of SVC obstruction may remain undiagnosed. In the present case, the presence of SVC thrombosis was purely coincidental. In fact, the obstruction was first noticed on diagnostic tests performed because of the malfunction of a totally implantable Porth a Cath placed into the superior vena cava (through right subclavian access), five years before, in a patient suffering from non-Hodgkin disease. Venography is the most appropriate diagnostic methodology which reveals the presence of a dilated azygos vein as a compensatory mechanism. Comparison with computed tomography allows to confirm the diagnosis and to identify the possible causes. Dilatation of the azygos vein, secondary to superior vena cava thrombosis, although a rare event, should be taken into consideration in those patients with CVC and who present with frequent episodes of deep venous thrombosis.

CASE REPORT

CASE REPORT

A 40 year-old man, diagnosed with centrilobular non Hodgkin lymphoma eight years prior to initial presentation was hospitalized in order to check the functionality of his totally implantable bicameral Porth a Cath (CVC radiopaque silicone 10 F DELTEC®, USA) that was placed by us in the superior vena cava three years after diagnosis. At the beginning of the disease, the clinical signs were characterised by superficial and deep lymphadenopathies and splenomegaly. Total body CT showed the presence of extensive cervical thoracic and abdominal lymphadenopathy.

Bone biopsy confirmed the presence of a medullary involvement with hemocytometric analysis showing: WBC 14750/mm³ (N 54%, L 41%, M 5%), Hb 13.9g/dl, PLT 285000/mm³.

Thereafter, a splenectomy was performed with its histological and immunohistological assessment confirming the diagnosis of a non-Hodgkin's disease of a probable centrilobular origin. Two years after diagnosis the patient developed a venous thrombosis of the superior cava vein, efficiently treated with Heparin therapy. Three years after diagnosis a central venous catheter (totally implantable bicameral Porth a Cath) was placed for chemotherapy, through

a right subclavian access and with its tip positioned in the superior vena cava (distal third).

In the same year, the patient developed a deep right subclavian-axillary-brachial vein thrombosis, which required another Heparin treatment.

One year after Porth a Cath placement, after further progression of the disease, the patient was submitted to a polychemotherapy program followed by a therapy cycle with monoclonal antibody anti-CD20.

The patient responded excellent to the treatment, and he proceeded to radiochemotherapy,

Five years after Porth a Cath placement (2005) the patient came back to assess the function of the central venous catheter which was not able to aspirate.

At the first radiological visit, an anomalous positioning of the catheter tip was noted on the frontal radiograph, which had a curvilinear displacement with right lateral concavity. The distal tip of the catheter was positioned at the level of the proximal third of the superior vena cava (Fig.1).

In order to better understand the case, a cavography was performed, which showed a stenosis of the superior vena cava at the level of the azygos vein exit that resulted in significant compensatory dilatation of the azygos vein. The calibre of the vena cava below the stenosis was reduced to 60%: 6mm (Fig. 2).

To better define the aetiology of this process, a contrast enhanced CT thorax and abdomen was performed and compared to a previous CT thorax obtained three years before (2002).

The findings were not present on the comparison study, excluding a congenital nature of this process. The azygos vein and the SVC had a normal calibre on the comparison study from 2002, whilst the CT from 2005 revealed the presence of an azygos considerably dilated (15 mm) in spite of a SVC that had a calibre considerably reduced (Fig. 3, Fig. 4).

The stenosis found in our patient at the level of the azygos vein exit, was therefore attributed mainly to two possible factors: SVC thrombosis deriving from complications related to the positioning of the central venous catheter; or radio-induced stenosis as a consequence of radiotherapy treatment the patient underwent.

DISCUSSION

The diagnosis of acute superior vena cava obstruction is based on the evidence of peculiar symptoms and signs. Symptoms include dyspnoea, headache, epistaxis, visual disturbances, hoarseness, dizziness, syncope, tongue swelling and haemoptysis. Physical signs include swelling of the face and upper extremities, jugular venous distension, facial congestion, chest or shoulder swelling, distended thoracic veins, proptosis, laryngeal oedema, conjunctival injection, impaired visual activity, and Bernard Horner syndrome (1). The most common causes of SVC obstruction include bronchogenic carcinoma, thyroid gland enlargement, mediastinitis and fibrosis secondary to histoplasmosis and tuberculosis, other intrathoracic mass lesions, Behcet's disease, and thrombosis and fibrosis following the insertion of central venous catheters (2). Thrombosis occurs in up to 40% of

patients with central catheters but only 1%-14% of these patients develop the SVC syndrome and the rate of development is estimated as 0.003%-0.2% for each day the catheter resides in the SVC (3).

When the thrombosis process develops gradually, the development of collateral venous pathways can make the diagnosis of superior vein cava stenosis difficult or even undetected.

These collateral pathways, bypassing the obstruction, provide venous drainage from the head, neck, chest, and upper extremities, and they empty into the azygos vein and the inferior vena cava. These pathways include: the internal thoracic-superior-inferior-epigastric veins on the left; the lateral thoracic-superficial epigastric veins on the right; the anterior and posterior intercostal veins; the accessory azygos, the hemiazygos, the jugular veins, the vertebral plexus, and the small tributaries of the thoracoabdominal wall and breast (1,2,3,4,5,6).

Venography is the most appropriate diagnostic methodology to confirm obstruction of the SVC. It allows to identify and classify four different patterns of venous collateral return: type I, partial obstruction (up to 90%) of the superior vena cava with patency of the azygos vein; type II, near-complete to complete obstruction (90-100%) of the superior vena cava with patency and antegrade flow through the azygos vein and into the right atrium; type III, near-complete to complete obstruction (90-100%) of the superior vena cava with reversal of azygos blood flow; type IV, complete obstruction of the superior vena cava and one or more of the major caval tributaries, including the azygos system (7).

Independently from the pathogenesis when stenosis II-IV developed, interventional radiology may play a role by performing fibrinolysis in acute obstruction and stent placement in case of chronic obstruction.

Contrast-enhanced computed tomography (CT) is a useful method in diagnosis of SVC occlusion. CT findings include an unopacified SVC with opacification of collateral vessels, whose presence is an accurate predictor of SVC occlusion. Intraluminal defects of the SVC may be caused by a thrombus, a stenosis due to multiple central line insertions or tumor invasion. Extraluminal compression is most commonly caused by mediastinal tumors or lymph nodes. If the tissue surrounding the occlusion is enhanced by contrast and the stenotic process involves a long tract of the vein the most probable diagnosis is a neoplastic process. Otherwise if the tissue is not enhanced by contrast and the stenotic process is short and with smooth surface the most reliable origin is a benign compression.

Usually in case of neoplastic causes type I patients are best managed by radiation and chemotherapy (bendamustine only) to reduce the neoplastic mass and do not require operation or bypass. SVC Types II to IV patients are treated by operation when symptoms of dyspnoea or cerebral venous hypertension are present. Type III patients are considered ideal for operation because the left brachiocephalic vein is usually available for bypass. Type IV patients may also be considered for operation, but it could be very difficult and may require venous thrombectomy (8).

The cause of the SVC stenosis in our case was thought to be due to CVC malposition, a known etiology of CVC

stenosis (9,10,11). There is a close association between the catheter tip position and thrombotic complications (12,13).

Our conclusion accords to several studies that have shown that the complication rate is lower when the catheter tip is positioned within the lower third of the superior vena cava or close to the junction of the SVC and the right atrium (14,15)

Besides, CVC distal catheter tip positioning high in the SVC seems to be the sole factor that was statistically predictive of malfunctions (16).

In the present clinical case, the distal tip of the catheter was positioned above the aforementioned level (SVC top half) showing a curvilinear displacement with a right lateral concavity which, presumably, produced irritation against the vessel wall, subsequent endothelial injury and the development of thrombosis (17).

It should be also mentioned the higher association between malignancies and the development of thrombosis.^{3, 11} The combination of these two factors, central venous catheter malpositioning and malignancy, is often responsible for the onset of thrombotic complications.

The infusion of chemotherapeutic agents may also cause progressive irritation of the vessel wall with injury to the endothelium and the development of thrombosis.

The malfunction of a CVC may be reduced by correctly placing its tip as close as possible to the SVC-right atrium junction. The correct positioning should be confirmed by fluoroscopy and radiography (Fig.1).

Last but not the least hypotheses to be considered is the possibility of a radiation-induced stenosis of the vessel lumen. Although arterial stenosis secondary to irradiation is more frequent, venous thrombosis and the subsequent luminal obstruction are also known sequela (14,17).

A condition that might resemble an acquired SVC obstruction is the duplication of inferior vena cava (incidence 0.2-3.0% in literature). In literature six cases of IVC duplication associated with a clinically relevant deep venous thrombosis are reported but the correlation between the duplication and the thrombosis is purely incidental and the development of the thrombosis involves lower extremities (18).

We conclude that it is rather rare (3) to find clinical cases of acquired compensatory dilatation of the azygos vein, similar to the one discussed in the current paper. Nevertheless, it is very important to consider such a diagnosis, especially in those patients with central venous catheters, malignancy and episodes of deep venous thrombosis.

TEACHING POINT

Recognition of dilated azygos vein, secondary to superior vena cava thrombosis should always be taken into consideration in those patients with central venous catheters and neoplasm. The tip of the central catheter should be positioned within the lower third of the superior vena cava (SVC) to lower the risk of catheter induced SVC thrombosis.

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FIGURES

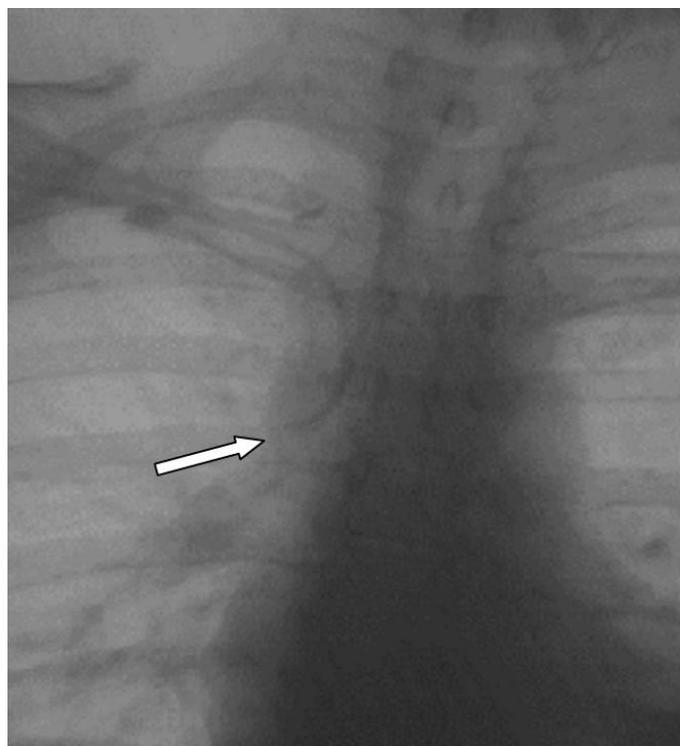


Figure 1. 40 year old man with compensatory dilatation of the azygos venous system secondary to superior vena cava occlusion. Fluoroscopic image of the chest in P-A projection demonstrates a curvilinear displacement of the central venous catheter with right lateral concavity. The distal tip of the catheter is positioned at the level of the proximal third of the superior vena cava (arrow).

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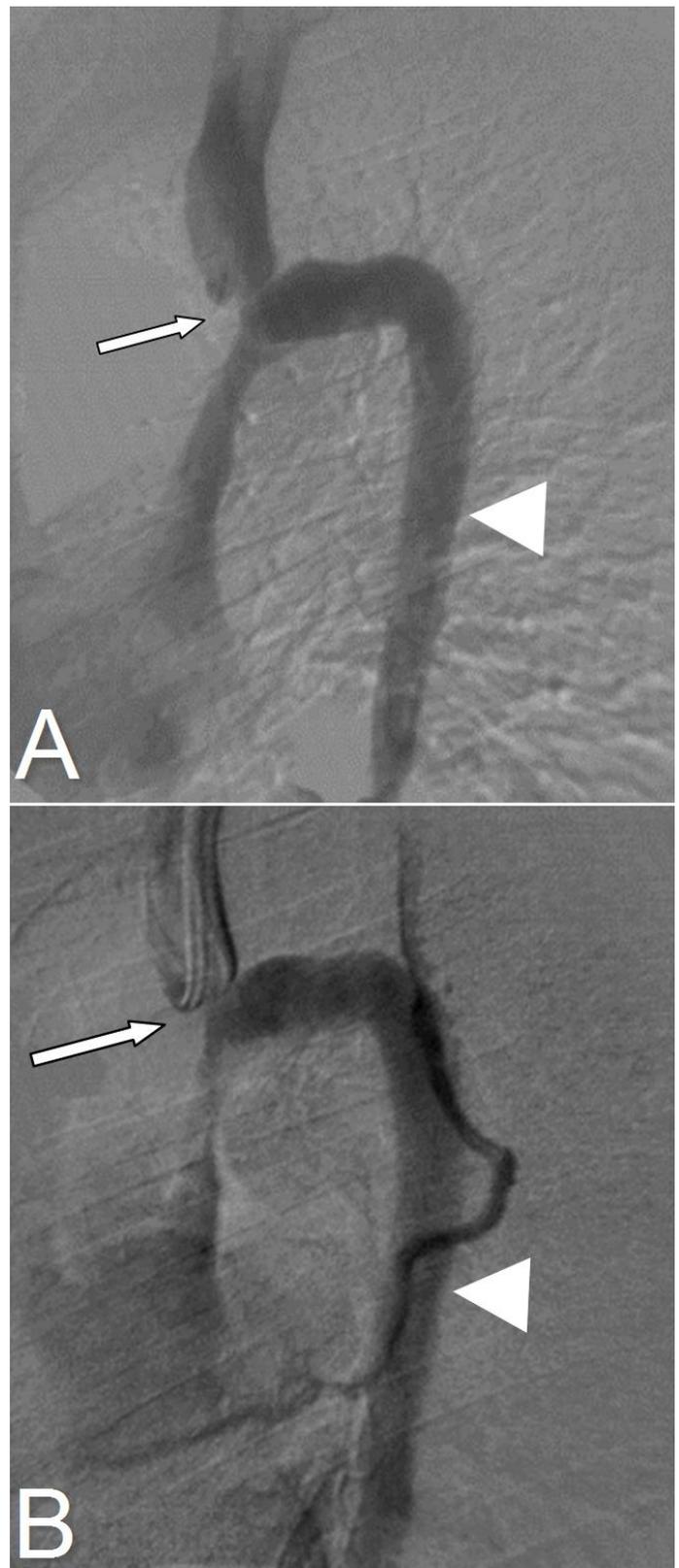


Figure 2. 2-a + 2-b: 40 year old man with compensatory dilatation of the azygos venous system secondary to superior vena cava occlusion. Cavography after contrast injection in lateral projection. Seen is left SVC stenosis at the level of the azygos vein exit. Please note the abrupt calibre change of the SVC (arrow) and compensatory hypertrophy of the azygos vein (arrowhead).

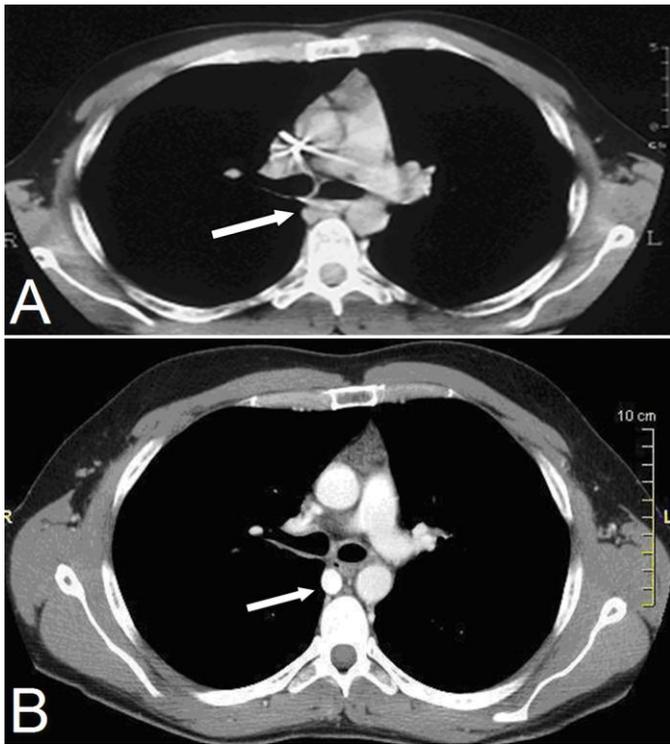


Figure 3. 40 year old man with compensatory dilatation of the azygos venous system secondary to superior vena cava occlusion. Contrast enhanced chest CT at the level of the carina. Comparison between the study from 2002 (3a) and 2005 (3b) demonstrates the presence of a hypertrophic azygos vein in the 2005 image (arrow).

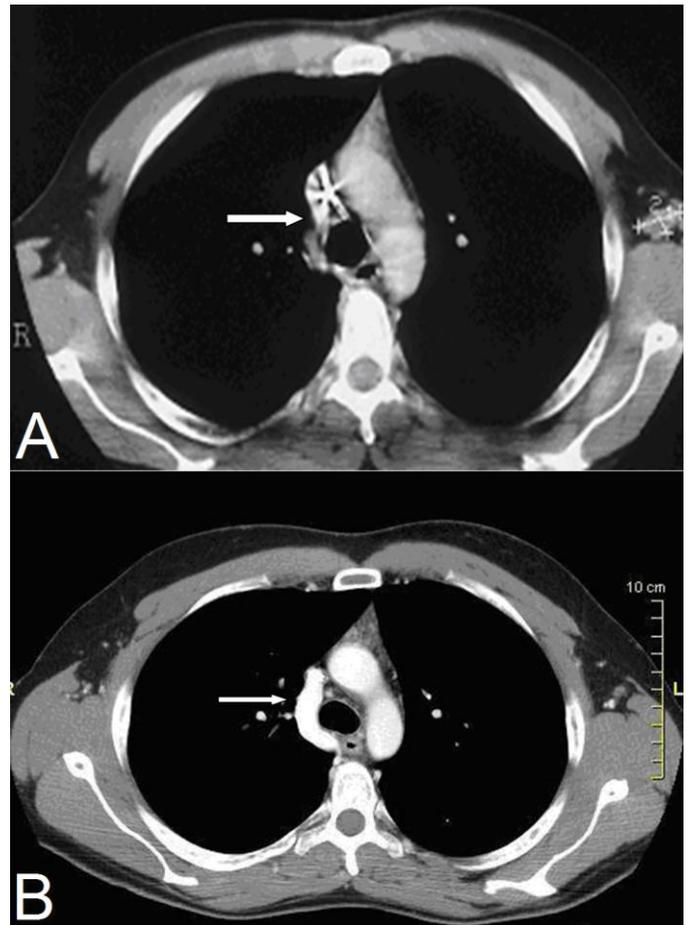


Figure 4. 40 year old man with compensatory dilatation of the azygos venous system secondary to superior vena cava occlusion. Contrast enhanced chest CT at the level of the azygos vein exit into the SVC. Compared to the study obtained in 2002 (4a), marked hypertrophy of the azygos vein arch (arrow) is noted on the image from 2005 (4b).

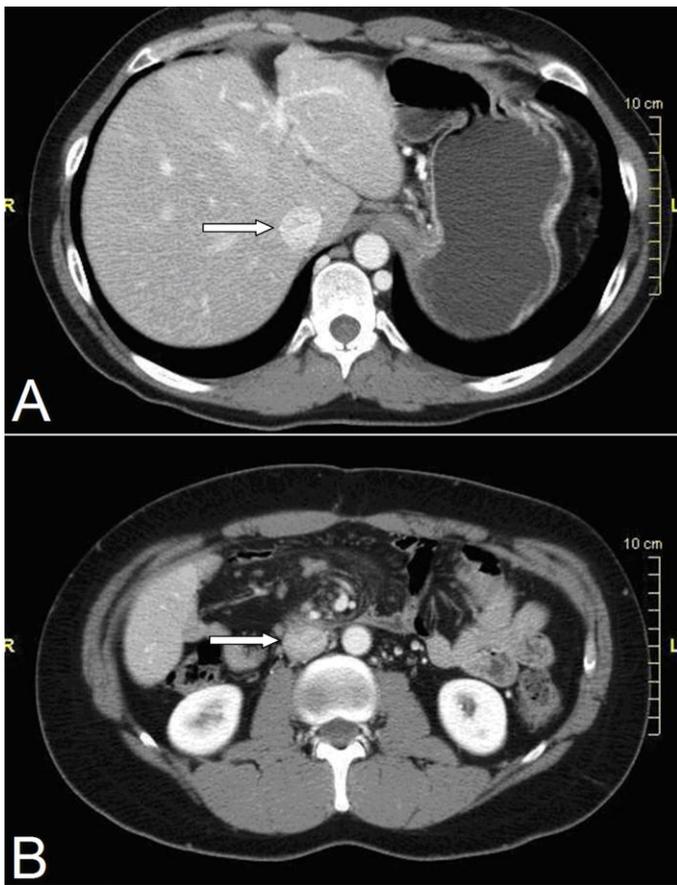


Figure 5. 40 year old man with compensatory dilatation of the azygos venous system secondary to superior vena cava occlusion. Contrast enhanced CT at the level of abdomen show normal inferior vena cava at intrahepatic (5a) and juxtarenal (5b) level without evidence of duplication of inferior vena cava.

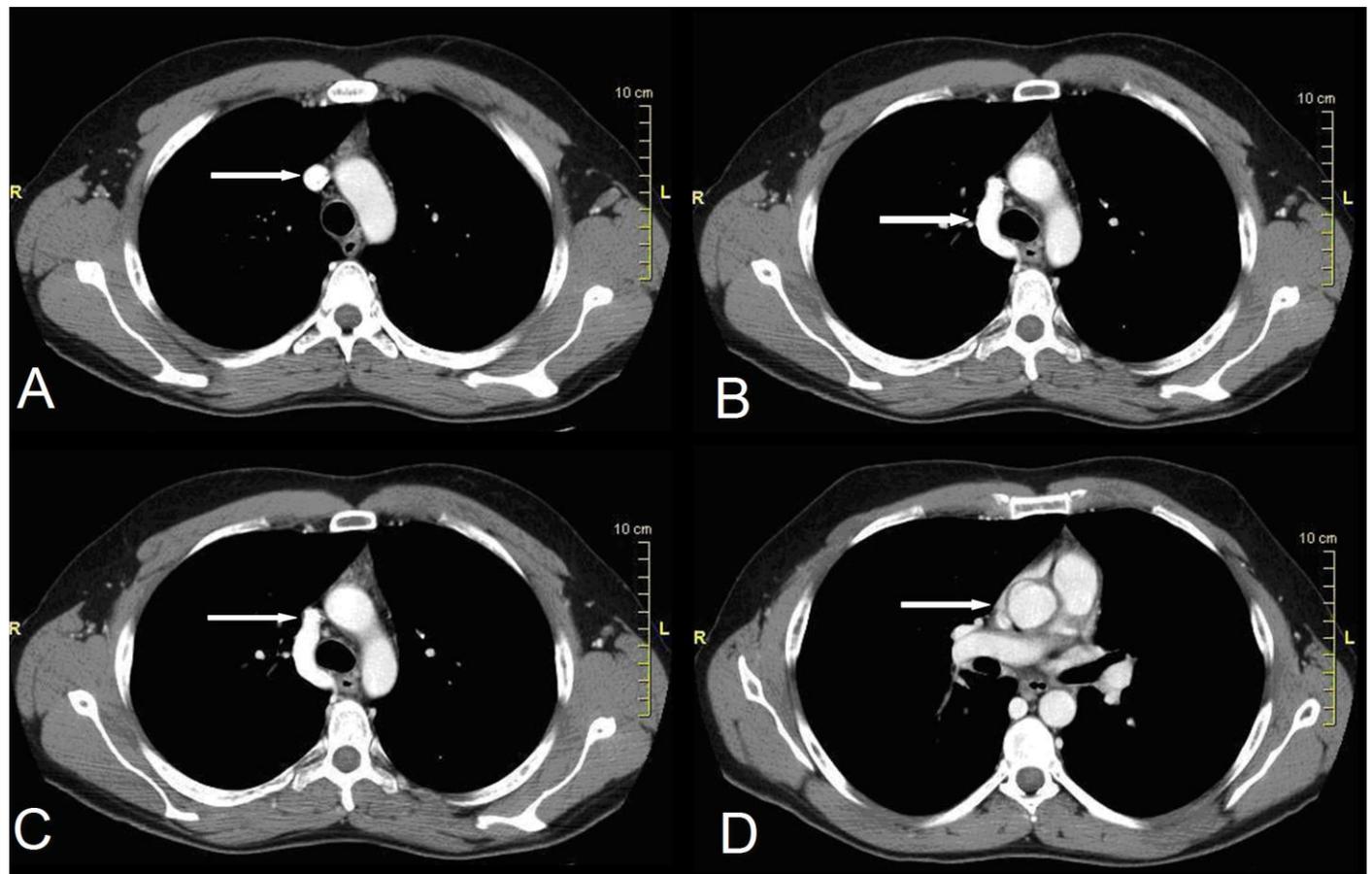


Figure 6. 40 year old man with compensatory dilatation of the azygos venous system secondary to superior vena cava occlusion. Contrast enhanced CT of the supra-stenotic tract of SVC (6a), at the level of the arch of the azygos vein (6b), at the level of the stenosis of SVC (6c) and at the sub-stenotic level (6d).

ABBREVIATIONS

SVC = Superior vena cava
CVC = Central venous catheter
WBC = white blood cells
Hb = Haemoglobin
PLT = Platelets

KEYWORDS

Azygos Vein, Superior Vena Cava, Central Venous Catheterization

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