Radio-anatomy of the superior vena cava syndrome and therapeutic orientations

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KEYWORDS
Superior vena cava syndrome; Phleboscanner; Endoprosthesis; Overload syndrome; Central catheter

Abstract Superior vena cava syndrome (SVCS) groups all the signs secondary to the obstruction of superior vena cava drainage and the increase in the venous pressure in the territories upstream. There are two major causes of SVCS: malignant, dominated by bronchopulmonary cancer, and benign, often secondary to the presence of poorly positioned implantable venous devices. CT scan is the key examination for the exploration of SVCS. It specifies the characteristics of the stenosis, its aetiology and detects collateral venous routes. Scannography reconstructions provide a true map of the obstacle, indispensable in planning the endovascular treatment.

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SVCS groups all the signs secondary to the obstruction of SVC and the increase in the venous pressure in the territories upstream. Malignant aetiologies are most common (74 to 95% of the cases) (Fig. 1), dominated by bronchopulmonary cancer (85% of the cases) [1,2]. It most often consists of small cell cancers [3]. The lymphomas are the second leading malignant aetiology (about 12% of the cases) [1]. Benign aetiologies are less common (3 to 20% of the cases) [1]. It often involves a thrombus around a central catheter, much more

Abbreviations: SVCS, Superior Vena Cava Syndrome; SVC, Superior Vena Cava; IVC, Inferior Vena Cava; MIP, Maximum Intensity Projection; ITV, Internal Thoracic Vein.

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Bronchopulmonary tumour responsible for a superior vena cava syndrome. The CT scan (coronal reconstruction) detects the tumoral mass (tip of arrow) provoking a focal occlusion of the superior vena cava (SVC) (arrow). The underlying SVC (wide arrow) is permeable and opacified by the collateral veins (re-entry by the azygos vein). Stasis of the contrast product in the SVC underlying the obstacle.

rarely around the wires of a pacemaker [4,5]. This thrombosis is favoured by the overly short positioning of the catheter in the SVC (Figs. 2–4) [4,6–8]. A central catheter is in normal position when the distal end is located at the entrance to the right atrium. An overly short catheter, whose distal end is located opposite venous convergences leading into the arch of the azygos vein at the convergence of the innominate veins (formerly called brachiocephalic veins), may come up against the vein walls and induce endothelial lesions by mechanical friction on the one hand, or may provoke rheological modifications in these zones of flow turbulence on the other hand. The other favouring factors are represented by the hypercoagulability of the cancer patient and the toxicity of the chemotherapy products injected by catheter [4]. The other benign causes of SVCS are rarer. It most often consists

Indirect azygos venous shunts (superior vena cava [SVC]-inferior vena cava [IVC] anastomoses) during a superior vena cava syndrome secondary to a poorly positioned catheter (too short). The diagram and the scan demonstrate the shunting by the pericardio-phenic vein. The latter drains in the bronchial, oesophageal and diaphragmatic veins that then join the azygos system. There are anastomoses between the bronchial and the pulmonary veins and between the oesophageal/diaphragmatic veins and the portal system.
Figure 4. Superior vena cava syndrome in a patient presenting a poorly positioned catheter. a: scan with injection, frontal reconstruction revealing an overly short catheter, responsible for thrombosis with partial obstruction of the superior vena cava (SVC) (arrow). b: insertion of an endoprosthesis in the SVC. c: enhanced CT scan (coronal section in maximum intensity projection [MIP]) revealing the good position of the prosthesis and its permeability (wide arrow).

Figure 5. Hugues Stovin syndrome. Superior vena cava syndrome during Behcet’s disease. Presence of pulmonary arterial aneurisms. Tight stenosis of the right innominate vein and the sub-azygos superior vena cava (tip of arrow). Spinal shunting (wide arrow) draining in the superior intercostal veins, then in the arch of the large azygos vein (white arrows) to join the sub-azygos superior vena cava (black arrows).
of extrinsic compressions: mediastinal haematoma, benign tumours (diving goitre, bronchogenic cyst, teratoma), acute or chronic mediastinitis, cardiovascular causes (aortic dissection or aneurism, constrictive pericarditis, pericardial effusion, atrial myxoma, etc.). Behcet’s disease may be responsible for thrombosis of the SVC and, when associated with pulmonary arterial aneurism, give rise to Hughes Stovin syndrome [9,10] (Fig. 5).

**Imaging**

The thoracic radiography may show signs of the development of collateral circulation: opacity above the right stem bronchus related to a dilation of the arch of the azygos, sub-aortic opacity or “aortic nipple” corresponding to the dilation of the left superior intercostal vein [11] (Fig. 2). The radiography may indicate the aetiology of SVCS: for example, tumoral mass or implantable venous device with a short catheter [2]. The SVC permeability may be evaluated by the analysis of four Doppler spectra of the subclavicular and internal jugular veins [12]. The reversal of the flow of the collateral veins attests to the steal syndrome of the SVC: the ITV has the same Doppler colour as its satellite artery (Fig. 6) [13]. The elimination of these anomalies is used to assess the efficacy of the treatment, in particular endovascular, of the SVCS. The scan is the key examination in the diagnosis and therapeutic strategy. The MRI may be an alternative, although this is a long examination, difficult to use in often dyspnoeic patients not able to bear the dorsal decubitus position.

The phleboscanner specifies the characteristics of the SVC obstruction and its aetiology: impairment of the vein wall (post-radiation stenosis, friction of an overly short central catheter), extrinsic compression (adenomegalies, neighbouring tumour) and cruric thrombosis (venous stasis, hypercoagulability). In our practice, the optimum study of the SVC requires a thoraco-abdomino-pelvic acquisition 80 seconds after the injection of 1.5 mL/kg of a non-ionic iodine contrast medium (300 to 400 mgI/mL) at the rate of 2 mL/s. This acquisition, at the vascular equilibrium phase, allows for homogenous venous opacification and avoids the false images of thrombus, generated by the flow artefacts observed on the early acquisitions. However, an early thoraco-abdomino-pelvic acquisition, obtained 15 seconds after injection, reveals all of the venous shunting. This acquisition is not indispensable in everyday practice although it does allow for the exhaustive study of the venous anatomy, the purpose of this paper.

Stanford’s classification, modified and adapted for the scanner by Qanadli, is used to quantify the degree of venous obstruction during a superior vena cava syndrome.

| Stage I | Stenosis < 90% of the superior vena cava |
| Stage II | 90 to 99% stenosis of the superior vena cava |
| Stage III | Occlusion of the superior vena cava |
| Stage IV | Occlusion of the superior vena cava and one or several of its tributaries |

**Table 1** Kishi score [26]. The Kishi score is used to quantify the clinical gravity of the superior vena cava syndrome. A score exceeding 4 is an indication for the insertion of a superior vena cava endoprosthesis by percutaneous route.

<table>
<thead>
<tr>
<th>Clinical signs</th>
<th>Weighting</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Neurological signs</strong></td>
<td></td>
</tr>
<tr>
<td>Awareness disorders, coma</td>
<td>4</td>
</tr>
<tr>
<td>Visual disorders, headache, vertigo, memory disorders</td>
<td>3</td>
</tr>
<tr>
<td>Mental disorders</td>
<td>2</td>
</tr>
<tr>
<td>Malaise</td>
<td>1</td>
</tr>
<tr>
<td><strong>Thoracic/pharyngeal-laryngeal signs</strong></td>
<td></td>
</tr>
<tr>
<td>Orthopnoea, laryngeal oedema</td>
<td>3</td>
</tr>
<tr>
<td>Stridor, dysphagia, dyspnoea</td>
<td>2</td>
</tr>
<tr>
<td>Coughing, pleuresy</td>
<td>1</td>
</tr>
<tr>
<td><strong>Facial signs</strong></td>
<td></td>
</tr>
<tr>
<td>Lip oedema, nasal obstruction, epistaxis</td>
<td>2</td>
</tr>
<tr>
<td>Facial oedema</td>
<td>1</td>
</tr>
<tr>
<td><strong>Vessel dilation (neck, face, arms)</strong></td>
<td>1</td>
</tr>
</tbody>
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**Table 2** CT classification of superior vena cava syndrome according to Qanadli [15]. It is used to quantify the degree of venous obstruction during a superior vena cava syndrome.
anastomosis between the superior and IVC system (Fig. 7). The azygos system consists of the large azygos vein, on the right, and the two small azygos veins, on the left [16]. Two situations can be distinguished according to the topography of the occlusion, below or above the arch of the large azygos vein. The first situation corresponds to a SVC occlusion located below the junction of the arch of the azygos vein. Shunting by the azygos system may occur directly or indirectly.

With direct azygos shunting, the blood flows counter-current in the azygos system and joins the IVC either by means of the internal roots of the azygos veins or by means of the ascending lumbar veins and then the external iliac veins (Fig. 7) [16]. The blood flow may also join the ITVs by means of the intercostal veins (Fig. 8). The ITVs are anastomosed to the epigastric and superficial epigastric veins that join the IVC system by means of the external iliac veins and the internal saphenous veins respectively (Fig. 9) [17].

With indirect azygos shuntings, the blood flows by the pericardo-phrenic veins. The latter communicate through their lower part with the oesophageal veins, the diaphragmatic veins, the mediastinal veins and the bronchial veins that join the azygos system and then the IVC system (Fig. 3) [16]. The large and small azygos veins communicate behind by the intra- and extra-spinal venous plexuses, allowing for transverse contralateral shunting (Fig. 9) [16].

The superficial venous system also creates a cavo-caval anastomosis. It is possible to distinguish the anastomoses between the innominate veins and the external iliac veins through the ITVs and the epigastric veins, and the anastomoses between the axillary veins and the internal saphenous veins through the external thoracic veins and the superficial epigastric veins (Fig. 10) [17]. These parietal veins are mutually anastomosed and may join the para-umbilical vein (Figs. 8 and 11) [17].

Other shunting to the pulmonary veins forms a right-left venous shunt. In fact, the bronchial veins are anastomosed in proximity with the azygos system, and distally with the pulmonary veins (Fig. 3) [16,18,19]. Direct anastomoses
Figure 9. Spinal anastomoses. These anastomoses enable the shunting towards the contralateral side, via the intra and extra-spinal plexuses.

Figure 10. Superficial venous anastomoses. The parietal (internal and external thoracic, epigastric and superficial epigastric) veins ensure the shunting between the inferior vena cava (IVC) and the superior vena cava (SVC).

between the oesophageal and pulmonary veins have also been described [20].

Other shuntings, towards the portal system, have been described. The inferior oesophageal veins anastomose with the left gastric vein [16]. The pericardial, diaphragmatic and parietal veins shunt the blood flow to the para-umbilical veins at the notch of the ligamentum teres [17]. This shunting to the para-umbilical vein accounts for the very intense enhancement of contrast medium observed opposite the groove of the ligamentum teres during SVCS. They are called "hot spot signs" (Figs. 8 and 11) [21–24]. The blood flow then joins the left portal branch at the rex recess [23,24] or the hepatic veins directly (Figs. 8 and 11).

The second situation corresponds to a SVC occlusion located above the junction of the arch of the azygos vein or at the occlusion of the innominate vein. In this situation, supra-ternal venous anastomoses develop, allowing for a transverse contralateral shunt (Fig. 12). Therefore, clinical SVCS is not generally observed if only one of the

Figure 11. Hepatic shunting (hot spot sign) during a superior vena cava syndrome. Injected scan (sagittal reconstruction in maximum intensity projection [MIP]) revealing dilated parietal veins draining in hepatic segment IV (determining an intense enhancement) then in the hepatic veins (black arrow) and the left portal branch (white arrow).

Figure 12. Superior vena cava syndrome by occlusion of the superior vena cava (SVC) upstream from the junction of the azygos vein. Supra-ternal anastomoses via the jugular and thyroid veins.
Innominate veins is occluded. The anterior jugular veins, leading into the innominate vein or the external jugular veins are directly or indirectly anastomosed by way of the thyroid veins. The thyroid veins also directly ensure the contralateral shunt of the blood flow [16]. Posterior anastomoses also exist by way of the vertebral veins and the spinal plexuses (Fig. 9) [16]. The blood joins the superior intercostal veins, the aygos veins and the SVC below the obstacle. Moreover, the blood flow can borrow the pericardial-phenic veins and the shunts of the aforementioned superficial parietal veins (Figs. 3 and 10).

**Symptoms**

The clinical signs are related to the increase in venous pressure upstream from the SVC obstacle and the development of venous shunts (Fig. 13). A so-called “pilgrim oedema” of the upper part of the thorax is observed with filling of the sub-clavicular hollows, oedema of the front (precocious palpebral oedema), cyanosis, dilatation of the superficial veins and swelling of the jugular veins [25]. Deep down, the oedema may provoke dyspnoea and coughing by tracheobronchial impairment, dysphagia and dysphonia [2,25]. At the maximum, encephalic venous hypertension may provoke a brain oedema with headache (increased with anteflexion), confusion, epilepsy and coma [26]. Kishi proposed a score of the clinical gravity in which the dyspnoea and neurological signs are pejorative [26]. The intensity of the symptomatology depends on the development of the collateral. Therefore, an acute SVCS is often much more poorly tolerated than a chronically evolving vena cava syndrome. In the latter case, the collaterality has the time to develop. Moreover, Stanford’s classification is also of prognostic value. In fact 80% of all patients with SVCS presenting respiratory distress or neurological signs have a Stanford type III or IV [27].

**Care**

The care is pluridisciplinary. The choice of the different treatments depends on the degree of urgency to treat the SVCS, the chemosensitivity of the cancer involved, and the scanner results [28]. As far as possible, a treatment should not be prescribed before obtaining a histology [29]. Medical care, including a semi-sitting position favours venous drainage. An effective dose of anticoagulant treatment and corticotherapy are usually prescribed. However, the efficacy of corticotherapy has not been demonstrated [3,29].

In case the histology is predictive of a good tumoral response (small cell carcinoma, lymphoma and germ cell tumours), the chemotherapy may induce the rapid improvement of the symptoms [28]. Similarly, external radiotherapy may be effective in 2 to 3 weeks [30,31]. However, in case of a recurrence, the vena cava syndrome may be poorly tolerated due to the occlusion of the collaterals, secondary to post-radiation fibrosis or a thrombosis. The association of radiotherapy and chemotherapy has not demonstrated any additional efficacy [28]. However, chemotherapy and irradiation have a longer delay before action than that of percutaneous radiology procedure and, in spite of the good initial results, the vena cava syndrome recurs in 10 to 32% of the cases [30]. In the acute thromboses, treatment by fibrinolysis has been proposed with the potential risk of bleeding of infra-clinical cerebral and bronchial metastases [32]. Bypass surgery with venous graft, maintaining several indications in benign SVCS [33] is advantageously replaced by endovascular treatment [34]. The original surgical indication during malignant SVCS is thymoma, before external irradiation and after adjuvant chemotherapy [35]. The second indication is thyroid cancer. In this case, a triple treatment associating surgery, external irradiation and iratherapy increases patient survival [36,37].

First described in 1986, the insertion of a SVC endoprosthesis is currently becoming the choice treatment for SVCS (non thrombotic venous axes or venous axes opened by angioplasty) (Figs. 4 and 13) [38]. It may be associated with the other means of treatment described above. Several studies have demonstrated its rapidity of action, efficacy and low morbidity [39–43]. Two families of endoprostheses can be distinguished: expandable endoprostheses requiring the inflation of a balloon on which the stent has previously been fixed and self-expandable endoprostheses that open under the effect of their radial strength as soon as their sheath has been removed [4]. The scanner helps in planning the insertion of the endoprosthesis [15]. In practice, we use...
self-expandable endoprostheses whose calibre is adapted to the diameter of the healthy SVC, as measured by phleboscanner. The length of the prosthesis is chosen according to the extent of the diseased zone to cover, also assessed by scanner. Prostheses with a diameter ranging from 14 to 16 mm and a length from 60 to 90 mm are most often used.

In case of impairment of the two innominate veins or their convergence, unilateral recanalisation generally suffices due to the richness of the transverse anastomoses [44]. The side may then be chosen by applying the concept of internal jugular dominance, as assessed by CT scan or ultrasound. The opening of a SVC stenosis inducing a fast cardiac return of the third compartment (oedema) may generate an "overload syndrome" with pre-capillary pulmonary hypertension and pulmonary oedema (Fig. 14) [45]. This complication is foreseen by the assessment of the cardiac function, a diuretic treatment and screened by short monitoring in an intensive care unit. The permeability of SVC prostheses seems to be satisfactory on a medium term basis [46]. The CT scan helps specify the characteristics of the stenosis (nature, location, extent, shunt routes, etc.) and plan for the insertion of the endoprosthesis [15]. In post-procedure, an anticoagulant treatment (heparin and relay by antivitamin K) is prescribed for 2 months. A phleboscanner carried out after 1 month is indispensable for the assessment of the tumoral response, the control of the permeability and prosthetic positioning and the disappearance of the collaterals (Fig. 4). It also enables the diagnosis of the complications [47]. The Doppler ultrasound remains an incomplete but simple way to assess the removal of the obstacle (Fig. 6). The medium term permeability of SVC prostheses seems to be acquired [46].

**Conclusion**

Mediastinal venous radio-anatomy may be easily explored in the presence of a SVCS leading to a number of shunt pathways. The phleboscanner, first intention examination when faced with a SVCS, specifies the reliability of the characteristics (location, extent, type, mediastinal environment) of the obstruction and allows for the precise planning of the endovascular treatment.

**References**


