A CASE FOR EDUCATION

Benign vena cava superior syndrome in patients with cardiac implantable electronic devices: Presentation and management

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Introduction

Venous occlusion that affects the subclavian or innominate vein is a common problem in cardiac implantable electronic device (CIED) patients, with a reported range between 3% and 5%. 1 In contrast, occlusion of the superior vena cava (SVC) not associated with malignancy (thus termed “benign”) is a rare condition (incidence 0.03%-0.15%) that causes a clinical syndrome of upper extremity and face swelling, pleural effusions, orthostatic hypotension, and collateral circulation and might even give rise to life-threatening complications if undetected, as illustrated by this case report. 2,3

Case report

An 84-year-old female patient was admitted to the surgery department of our hospital for the operation of a symptomatic anal prolapse. Past medical history included a dual-chamber pacemaker implantation in 2003 owing to symptomatic second-degree atrioventricular block, with generator change in 2017 owing to battery depletion. Left ventricular function was reported as previously normal with moderate mitral regurgitation. On examination, peripheral upper edema was noted. Pulmonary auscultation and percussion was consistent with bilateral pleural effusions, which were confirmed by chest radiograph (Figure 1). The patient was treated with oral loop diuretics and intestinal cleaning for surgery was initiated. After induction of anesthesia the patient was severely hypotensive. Vasopressors were given via a right jugular central line. As the patient did not respond, cardiac resuscitation had to be initiated. After further vasopressors and forced volume infusion the patient was stabilized. Emergency echocardiography showed good left ventricular function and no sign of right heart dysfunction. A moderate pericardial effusion was seen with possible hemodynamic compromise. Emergency pericardiocentesis was performed and 150 mL of amber-colored fluid was aspirated. Pulmonary embolism was ruled out by contrast computed tomography scan and significant right-sided pleural effusion was confirmed. A complete angiographic occlusion of the vena cava superior was diagnosed with collateral circulation via anterior thoracic veins and the vena azygos (Figure 2A–D). Pericardial veins were also dilated (Figure 2B). After stabilization the patient was extubated and transferred to an intermediate care ward. Pleural effusion was aspirated. Analysis of pleural fluid showed a protein of 2g/dL with an LDH of 250 U/L (serum LDH of 128 U/L) while pericardial effusion had a total protein of 4 g/dL with an LDL of 340 U/L. Serum protein levels were in the normal range. Triglycerides were not present in either effusion. Malignant and inflammatory causes of pleural and pericardial effusion were excluded. Dedicated echocardiography did not show a cardiac cause for pleural effusions; particularly significant diastolic dysfunction was excluded (Supplemental Figure 1). Pleural effusions had to be tapped repeatedly owing to recurrence despite optimal medical therapy including sequential
nephron blockade over several weeks with a range of LDH values between 130 and 360 U/L and a constant protein level of 2 g/dL. Further medical history revealed that the thoracic collateral circulation had been present for several months, supporting the assumption that the SVC occlusion was chronic. As the patient was symptomatic in terms of exertional dyspnea and orthostatic intolerance, recanalization of the chronic occlusion by balloon angioplasty ± stenting was scheduled.

The procedure was carried out under local anesthesia. A 10F sheath was inserted via the right common femoral vein. A hydrophilic guidewire was passed through the venous occlusion and 6F pigtail catheter was placed into the innominate vein. Manual digital subtraction angiography series were performed, confirming a total occlusion of the SVC beginning at the right atrium 5 cm in length and retrograde flow via the enlarged azygos vein (Figure 3A; Video 1). The occlusion was recanalized by incremental balloon angioplasty with 10 × 60 mm, 16 × 40 mm, and 18 × 40 mm balloons (Figure 3B; Video 2). Owing to significant elastic recoil (Figure 3C, Video 3), a self-expanding 20 × 60 mm uncovered stent (Optimed Sinus XL; Optimed, Esslingen, Germany) was placed into the SVC, followed by dilatation with an 18 × 40 mm balloon. Digital subtraction angiography confirmed an optimal angiographic result. Complete cessation of collateral blood flow via the azygos vein was confirmed (Figure 3D, Video 4). Oral anticoagulation with rivaroxaban 20 mg/d was reinstituted after the procedure. The subsequent clinical course was uneventful.

Figure 1  Chest radiograph with implanted dual-chamber pacemaker and bilateral pleural effusion.

Figure 2  Contrast thoracic computed tomography scan. Contrast medium was applied via a right cubital vein. A: Total occlusion of the superior vena cava 5 cm in length. The presumed remaining lumen is due to pacemaker wire artefact. B: Frontal plane showing extensive thoracic collaterals via the internal mammaria veins. Also note the contrasted pericardial vein. C: Reconstruction of thoracic collaterals. D: Retrograde filling of the azygos vein.
Impedances, sensing, and pacing thresholds of both leads were unchanged compared to preinterventional values. A follow-up visit after 6 weeks showed complete regression of collateral circulation with no signs of upper limb edema or pleural effusion. The patient’s dyspnea and orthostatic intolerance had completely disappeared. A stable clinical course was documented at 6-month follow-up (Supplemental Figure 2A/B).

**Discussion**

This case illustrates that the presence of recurrent pleural effusion, upper limb and face edema, and collateral thoracic
circulation in a CIED patient with endovascular leads should prompt further investigation. Collateral thoracic circulation might be mistaken for caput medusa associated with liver disease, which is more caudal and shows the typical signs and laboratory values of liver disease.

Several diagnostic and therapeutic steps have to be considered in the management of benign vena cava superior syndrome; venous phase-contrast computed tomography is the investigation of choice to determine a total or subtotal occlusion of the vena cava superior. Firstly, it has to be determined if the signs and symptoms are caused by the venous occlusion or if alternative causes for pleural effusion exist. Diagnostic and therapeutic aspiration of pleural effusions is definitely one of the first steps. The pathophysiological sequence for pleural and pericardial effusions is not entirely clear; however, increased intravascular hydrostatic pressure in the SVC with subsequent increased interstitial fluid in the lungs does definitely play a role.5 As such, it is suggestive that pleural effusions in benign vena cava superior syndrome are transudative; however, serial analysis of pleural effusions in our case could be classified as either transudative or exudative according to Light’s criteria. It must be taken into account that transudates might be “converted” to exudates following diuretic therapy.1 Presence of triglycerides might hint at the presence of chylothorax, which is associated with mediastinal fibrosis.3–4 Pericardial effusion is usually not described in conjunction with benign SVC syndrome; however, the dilated pericardial veins in our patient might be associated with increased intrapericardial pressures and subsequent effusion. Echocardiography can rule out significant valvular heart disease and systolic and diastolic dysfunction of the heart. Hypoproteinemia and autoimmune disease has to be ruled out as a cause for pleural effusions.

Secondly, if the patient’s symptoms are deemed to be related to vena cava occlusion, therapeutic strategy depends on the patient’s symptoms. If the patient remains symptomatic despite pleurocentesis and optimal diuretic therapy, interventional therapy may be warranted. Systems for the classification of severity and management of vena cava superior syndrome exist but may be of limited clinical value in CIED patients owing to the rapidly evolving therapeutic choices for endovascular treatment, lead extraction, and leadless pacing.5,6 Diminished cardiac reserve (postural syncope), upper limb edema with facial swelling, and refractory pulmonary effusion will usually require revascularization of venous occlusion.2,3 The clinical presentation of our case was exceptional, and intravascular hypovolemia was certainly exacerbated by the preoperative diuretic therapy and bowel preparation. Catecholamines applied via a central line will also have a delayed effect on cardiac inotropy in central SVC occlusion, which explains the initial refractoriness to intravenous inotropes following resuscitation.

Thirdly, if persisting severe symptoms require interventional therapy, the modality of interventional therapy (endovascular vs operative) needs to be determined. Thrombolysis may be considered in acute thrombotic occlusions, ie, in the first 2–4 weeks following device implantation. In all other patients endovascular recanalization is the treatment of choice in benign vena cava superior syndrome, with the exception of patients with fibrosing mediastinitis where extensive fibrosis and subsequent collateralization might occur.2,3,7 Reocclusion after endovascular treatment is also an option for surgical treatment.7

Finally, in CIED patients a strategy for how to deal with the implanted pacemaker / implantable cardioverter-defibrillator (ICD) leads has to be established. Percutaneous transluminal angioplasty only might be attempted in CIED patients to avoid jailing of pacemaker/ICD leads; however, significant elastic recoil is observed in the vast majority of cases requiring stent implantation, as in our patient.8 Some investigators argue that if the patient is to undergo stent deployment as treatment, lead removal should be performed prior to stent placement to avoid entrapment of the lead between the stent and the vessel wall; however, the additional risk of lead extraction has to be weighed against potential benefits.9 CIED infection is an absolute indication for lead extraction, while prior lead extraction should be considered in younger patients where future CIED infection with the subsequent necessity for complete system removal might be a potential risk. Lead extraction using laser sheaths or mechanical extraction tools provides at the same time vascular access for balloon angioplasty.10 These techniques might be combined with leadless pacemakers or subcutaneous ICD to reduce the risk of recurrent occlusion.10

Whether prolonged oral anticoagulation will prevent recurrent occlusion is not entirely clear; however, a recent study by Haddad and colleagues11 does not suggest a clinical benefit of prolonged oral anticoagulation after endovascular treatment.

Conclusion
Benign vena cava superior syndrome is an uncommon complication in CIED patients. Refractory pleural effusions, upper extremity and head and neck swelling, collateral thoracic circulation, and low cardiac output aggravated by orthostatic challenge may be presenting features, which should prompt further investigation. Endovascular treatment in symptomatic benign vena cava superior syndrome is an excellent treatment option that usually requires stent implantation. Entrapment of existing leads vs prior removal with subsequent reimplantation has to be discussed individually.

Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2020.06.018.

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