

Superior vena cava syndrome associated with implantable cardiac devices procedures (RCD code: VIII)

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Abstract

INTRODUCTION: Superior vena cava (SVC) syndrome is a rare clinical disorder associated with obstructing venous outflow through the SVC. Although the most common cause is mediastinal tumors, the growing number of implantable cardiac devices procedures has resulted in more patients with SVC syndrome of non-malignant etiology. **DISCUSSION:** It has been shown repeatedly that the use of cardiac implantable electronic devices (CIEDs) improves the quality of life of patients with symptomatic arrhythmias and reduces the mortality of patients at risk of sudden cardiac death, but the risk of complications also exists. **CONCLUSION:** The most common finding in patients with cardiac devices is asymptomatic venous thrombosis, but serious complications, including SVC obstruction should always be considered. The rapid identification of clinical symptoms and the implementation of appropriate measures are a key to preventing the morbidity and mortality of the patients. *JRCD* 2018; 4 (1): 5–8

Key words: rare disease, permanent pacemaker, implantable cardioverter-defibrillator, cardiac resynchronization therapy, multimodality imaging

Introduction

Pathogenesis

Superior vena cava (SVC) syndrome is the constellation of clinical symptoms resulting from the obstruction of blood flow through the SVC. The pathogenesis of the SVC occlusion is multifactorial, including catheter-induced endothelial injury/dysfunction with smooth muscle proliferation, abnormality of blood coagulation and an increased turbulent flow due to stenosis of vein's diameter by the catheter. SVC obstruction involves thrombosis, stenosis, or a combination of both and causes the increase in venous pressure above the obstacle.

Impairment of venous return to the heart from the head, neck, thorax and upper extremities is the reason for the collateral circulation development. Hence, the diagnosis is generally based on clinical signs and confirmed by imaging (radiography, computed tomographic angiography or venography).

Etiology

An estimated 15,000 cases of SVC syndrome occur each year in the United States ^[1], but the etiology has significantly changed, mostly due to the development of new invasive intravascular techniques. SVC syndrome was first described by William Hunter in 1757 in a patient with a syphilitic aortic aneurysm. Infectious causes, such as fibrosing mediastinitis from tuberculosis and thoracic aortic aneurysm from syphilis were the most common causes of this disease even until the fifties of the 20th century [1]. It can be assumed that this was because malignant tumors as the cause of the superior vena cava syndrome were not sufficiently diagnosed. Nowadays, most SVC syndromes are associated with advanced malignant diseases responsible for invasion of the venous intima or an extrinsic mass effect.

Lung, breast and mediastinal neoplasms account for more than 85% of all cases with the small-cell lung carcinoma being the most common cause [2].

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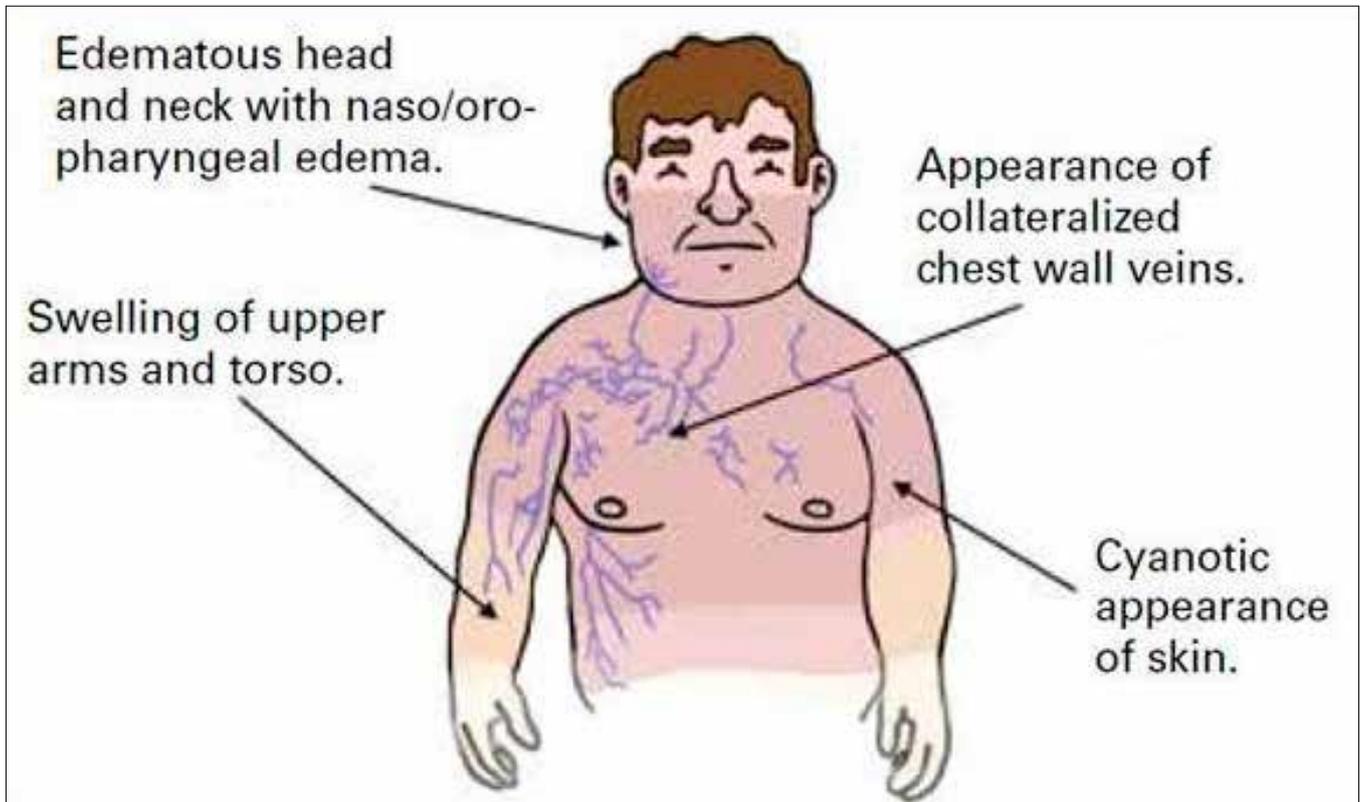


Figure 1. Symptoms of superior vena cava syndrome (SVCS)

Morbidity

Recently, with the increased use of intra-vascular devices, more cases have been observed after pacemaker/implantable cardioverter-defibrillator (ICD) implantations. Since the thrombosis and occlusion of SVC caused by the permanent transvenous cardiac pacing was described for the first time in 1973 by Wertheimer M, et al. [3], serious thromboembolic complications related to ICD and pacemaker leads have been reported in 0.6% to 3.5% of cases. ^[4] SVC syndrome is generally rare – the incidence of pacemaker/ICD-induced SVC syndrome has been occurred in less than 0,1% of patients [5].

Discussion

Cardiac implantable electronic devices (CIEDs) implantation is still growing. Since the transvenous pacing leads were used in 1960s for the first time, the treatment of arrhythmias and heart failure has significantly evolved. The 11th world survey of cardiac pacing and implantable cardioverter-defibrillators encompassing more than 80% of all the pacemakers and ICDs (in it cardiac resynchronization therapy devices – CRTs) implanted worldwide involved more than a million of pacemakers and about 330,000 ICDs (including CRTs) implanted during 2009 [6]. As the indications for implanting these devices are increasing, it may be assumed that the number of procedures related to CIEDs will enlarge annually and thus it is expected that complications will occur.

Complications associated with CIEDs

The risk of any complications following CIEDs implantation is approximately between 5% and 6% after pacemakers' implantation [7] and nearly 7,4% following ICDs/CRTs implantation procedures [8].

The minor complications in patients who underwent cardiac devices implantation procedures are described more often than the major ones with asymptomatic venous thrombosis being the most common finding [9].

As already mentioned, venous obstruction involves thrombosis, stenosis or combination of both. There is paucity of data on venous occlusion following device implantation. Moreover, it is challenging to analyze venous occlusions due to different definition of this disorder used in the literature. It was shown that various degrees of venous obstruction occur in up to 32% of patients receiving CIED [9, 10].

Risk factors for SVC syndrome development

In contrast to an incidence of between 8% and 21% of occlusion of the subclavian or brachiocephalic vein, the incidence of CIEDs-induced SVC stenosis is reported to be strikingly low, less than 0.1% [5, 10]. However, when it occurs, it is associated with significant mortality and adverse prognosis. Although no clear risk factors have been identified, several predictors of venous (including SVC) obstruction were identified – Table 1 [9, 11, 12].

It is not completely clear if congestive heart failure is associated with an increased risk, result can be related with protective effect of anticoagulant treatment prescribed for various reasons in patients

Table 1. Risk factors for CIEDs-induced venous obstruction [9, 11, 12]

	Increased Risk	Appropriate References
Patient data		
Sex	Female	[9]
Age(years)	>71.8	[9]
Body mass index(kg/m ²)	>27.9	[9]
Medical history		
History of venous thromboembolism	Yes	[9]
Active cancer	Yes	[9]
Major surgery/trauma	Yes	[9]
Upper limb paralysis	Yes	[9]
Hormone therapy	Yes	[9]
Acute myocardial infarction	Yes	[9]
Cardiac electronic devices history		
Device upgrade history	Yes	[9], [12]
Presence of multiple endocardial leads	Yes	[9], [12]
Previous use of temporary pacemaker	Yes	[9], [12]
Lead infection	Yes	[9]
Use of dual coil leads	Yes	[9]
Genetic predisposition		
Factor V Leiden/prothrombin G20210A	Yes	[9]
Factor VIII:C (IU/dL)	>205.5	[9]
Family history of venous thromboembolism	No	[9]
Drugs		
Lack of oral anticoagulant treatment	Yes	[9]
Periprocedural data		
Lead size	No	[11], [12]
Lead material	No	[11], [12]
Choice of access site	No	[11], [12]

with heart disease [9]. In turn, left ventricular ejection fraction (LVEF) \leq 40% was recognized as an independent risk factors to a higher incidence of venous stenosis or thrombosis[12]. No influence of family history of venous thrombosis on CIEDs-induced

venous obstruction can be explained by an inadequately collected medical history.

Surprisingly, neither the hardware (lead size and material) nor the vascular access (cephalic cut down, subclavian or axillary puncture) appears to affect rate of venous complications [11].

How can we recognize the symptoms of SVC syndrome?

Despite relatively high incidence of documented venous occlusion, most patients remain asymptomatic because of the development of an adequate venous collateral circulation. The symptoms are usually non-specific. Classic signs are very rare, however they are easily recognizable and allow to predict SVC obstruction with high probability. The limited patency of SVC increase the venous pressure in the upper body from the SVC obstruction resulting in edema of head and neck, naso/oropharyngeal, upper torso and arms.

The elevated hydrostatic pressure cause impediment of blood drainage from upper body what is visible as dilated veins – the most recognizable symptom of SVC syndrome. Consecutive signs of decreased venous flow are cyanosis and plethora (Figure 1). Subsequently, edema of the larynx or pharynx gradually cause stridor, dyspnea, dysphagia and cough. An important and one of the most characteristic clinical symptoms is the Horner syndrome caused by sympathetic trunk compression. Rarely, if venous return to the right atrium is completely blocked, patients can develop life-threatening complications such as hemodynamic collapse or cerebral edema, causing headache, confusion, dizziness or loss of consciousness.

Diagnosis and options of treatment of SVC syndrome

Diagnosis of SVC syndrome is based mainly on physical examination (the symptoms described above) and chest x-ray. Chest x-ray may be performed as a first test, but the most important radiological investigation is computed tomography (CT) of chest with intravenous contrast. It confirms the suspicion and shows the exact location, severity, and associated pathology. Magnetic resonance imaging (MRI) has no advantage over CT, except patients with renal failure. Ultrasound of the upper extremities is one of second-line examination, which is non-invasive and helps in identification of venous occlusion or thrombosis.

Sometimes bilateral upper-extremity venography can be useful, but nowadays, when CT and MRI are in common use, venography is not required for diagnosis.

Various options of treatment are nowadays used to manage the problem of CIEDs-induced venous occlusion include long-term anticoagulation, thrombolysis, percutaneous transluminal balloon venoplasty, endovascular stenting or finally surgical intervention.

Anticoagulants show an efficacy of up to 88% if they are included in the treatment within 5 days of diagnosis [13]. According to some authors, anticoagulants should be strictly used after an incidence of SVC obstruction caused by thrombus [14].

Although stenting is characterized by high efficiency and ensures quick resolution of clinical symptoms, it is not usually recommended as first-line of treatment in patients with non-malignant cause

of SVC obstruction and in patients with anticipated increased life expectancy, because stent occlusion can occur at any time [15].

Open surgery is the treatment of last choice, mainly reserved for symptomatic occlusion when the other methods have failed. The choice of treatment should be individualized to the patient and depends on the duration, extent, and site of venous occlusion as well as the accompanying symptoms.

Conclusions

With growing elderly population with more accompanying diseases and increasing number of CIED-s procedures performed, thromboembolic complications must be accounted. We would like to draw the attention of the clinicians to patients with any cardiac device, consideration of co-morbidities and risk factors for venous occlusion/stenosis during follow-up to better prevent and deal with this problem in the future.

Contribution statement

SJ contributed to research concept and design, collected the data and wrote the manuscript. LJ contributed to research concept and design, collected the data and revised the manuscript.

All authors contributed to the interpretation of the data, reviewed the manuscript, and approved its final version before submission.

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