

Superior vena cava syndrome associated with a pacemaker cable

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None

Superior vena cava syndrome (SVCS) is a rare disorder usually caused by a neoplasm, although other causes are involved in about 20% of cases. Such other causes are chronic fibrosing mediastinitis due to infection or radiation (42%-70%) and venous thrombosis (6%-27%). The prevalence of SVCS ranges from 0.03% to 0.4% but, because intravenous catheters (for pacemakers and defibrillators) are being implanted in a growing number of patients, the frequency is rising spectacularly. We report the case of a woman with SVCS related to a pacemaker cable. Interesting features of the case include the manner of detection and diagnosis. We emphasize the importance of physical appearance and the diagnostic and therapeutic measures to take. [Emergencias 2009;21:151-153]

Key words: Superior vena cava syndrome. Pacemaker.

Introduction

The superior vena cava (SVC) is a thin-walled vessel in the anterosuperior mediastinum whose function is venous drainage of the head, neck, upper extremities and an upper portion of the chest towards the right atrium. It is located near the trachea, sternum, right main bronchus, aorta and pulmonary artery, surrounded by parahilar and paratracheal lymph nodes. It may easily be compressed by any space-occupying lesion, resulting in superior vena cava syndrome (SVCS). SVC obstruction conditions a redistribution of venous flow towards the azygos, internal mammary, paraspinal, lateral oesophageal and subcutaneous veins.

SVCS was first described in 1757 William Hunter, in a patient with syphilitic aneurysm of the aorta. In 1973 Wertheimer et al published the first sporadic cases of SVCS in pacemaker carriers¹. It is currently a well-known complication, also associated with implanted cardiac defibrillators; however, appearance of the classic signs of SVCS is rare (0.2-3.3%). We present the case of a pacemaker carrier with a clinical picture compatible with SVCS.

Case report

This was a 65-year-old woman with a history of hypertension, obesity, depressive syndrome and peritonitis. She had received a permanent implanted pacemaker in 1994 for a bradycardia-tachycardia syndrome; due to subcutaneous pacemaker infection in 2005, the device was replaced but not all the cables of the first pacemaker were removed. Two months before this ED consultation, she suffered progressive swelling of the face and neck (Figure 1), two-pillow orthopnea, hoarseness, palpebral and facial oedema and ocular reddening which was more pronounced in the mornings but then improved. This was unsuccessfully treated as an allergic reaction; added to this, the patient had recently experienced dyspnoea in repose, which motivated her visit to our ED. Physical examination showed facial oedema, with bilateral conjunctival injection, pacemaker placement scars on the chest and collateral circulation in the pectoral and parasternal regions. Haemogram, blood tests and coagulation were normal. D-dimer was 0.98 mg/dL (normal < 0.5 mg/dL) and basal arterial gasometry showed: pH

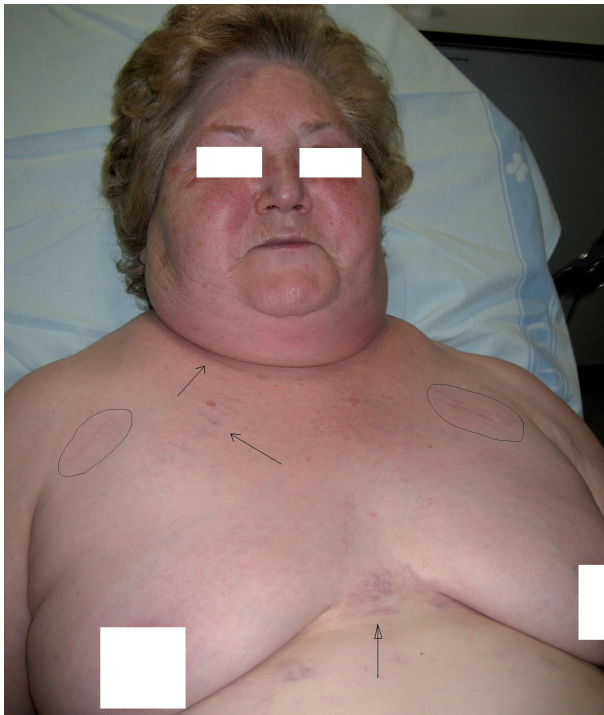


Figure 1. Image obtained on admission, showing facial and neck oedema, collateral circulation in the pectoral and xyphoid region (black arrows) as well as the pacemaker scars (circles).

7.38, pO₂ 65 mmHg, pCO₂ 45 mmHg and HCO₃ 26.6 mmol/L. CT angiography of the chest (Figures 2 and 3) revealed total VCS opacification, with multiple collateral veins, without evidence of pulmonary artery thrombosis. The patient was diagnosed with SVCS associated with pacemaker cables and was admitted to hospital for anticoagulation treatment.

Discussion

SVCS is an unusual condition usually associated with malignant neoplasia. However, in 20% of cases the etiology is not tumoral; of these, the most frequent causes include chronic fibrosing (infectious, post-radiation and idiopathic) (42-70%) and venous thrombosis (6-27%)². VCS stenosis or occlusion due to venous thrombosis is a well-known complication of permanent cardiac pacemaker insertion³⁻⁶; however, SVCS is rare, appearing in 1 of every 650-3,100 patients.

The time interval between pacemaker implantation and diagnosis of SVCS is 75 ± 66 months; in some asymptomatic cases this may be an incidental finding. Thus, thrombosis is only clinically significant as SVCS, pulmonary thromboembolism or sudden death in only 0.2-3.3% of all carriers.

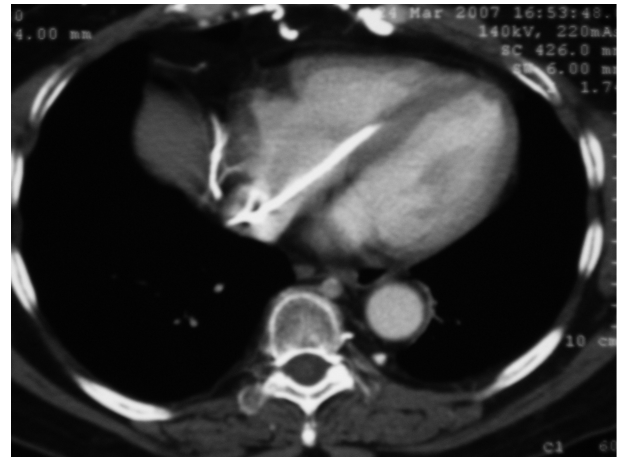


Figure 2. CT angiography showing the absence of filling in the superior vena cava and collateral filling.

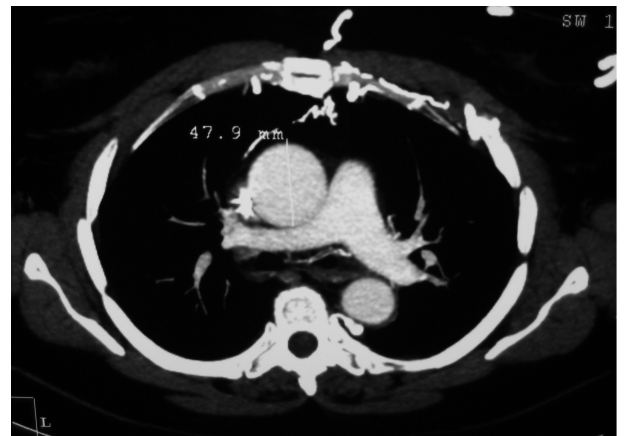


Figure 3. CT angiography showing the aorta and permeable pulmonary arteries as well as abundant collateral circulation including chest and subcutaneous branching.

Factors favouring venous thrombosis include intravenous catheter diameter (frequent in dialysis catheters), catheter material (improved tolerance of silicon compared to polyurethane catheters), patient condition and type of drugs administered (antimycotics, parenteral nutrition)^{7,8}.

Melzer et al⁷ reported an incidence of 1/360 patients (7/2,500) and predisposing factors were the presence of thrombophilia, multiple cables and/or partially extracted cables, infection and the use of polyurethane cables.

The underlying mechanism is fibrin deposition on the surface of the pacemaker points causing them to penetrate the vein, especially the particular points such as the confluence of the right atrial and the inferior vena cava⁷.

Symptoms of SVCS generally include facial and neck oedema, cough and dyspnoea; and occasionally cephalgia, tongue oedema, dizziness, epistaxis, conjunctival injection and rarely haemoptysis.

sis. Physical examination shows distension of the neck veins and collateral circulation of the chest wall. Horner syndrome may appear due to sympathetic chain compression in the mediastinum².

The diagnosis of SVCS is clinical, although radiology may also show mediastinal widening and pleural effusion, and the causal tumour may be identified. CT angiography is the diagnostic test of choice since it enables precise localization of the obstruction and can be used to guide the site of the biopsy. Cavography or venography can also be used.

Therapeutic management of SVCS is etiological, but in the case of pacemaker or catheter-induced thrombosis, treatment may involve anti-thrombolytics, with or without surgery, heparin with or without thrombolytics, angioplasty with or without thrombolytics and surgery^{9,10}. Anticoagulation with dicoumarines is an acceptable option in certain patients; however, due to the scarcity of long-term outcome studies, treatment duration is not clearly established and therefore should be individually decided¹⁰.

Other options include thrombectomy, endovascular prosthesis and stent implantation. Occasionally, surgery is needed to remove the pacemaker system, with venous revascularization and angioplasty and stent placement or implantation of a new pacemaker system¹⁰. Centella et al¹¹ have recently reported the use of percutaneous techniques to extract faulty pacemaker and defibrillator electrodes, with a success rate of 96.8%, using

a combination of different methods which avoid the need for more invasive surgery. Since these techniques also involve some major complications (2.5%), they should be performed by experienced teams.

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Síndrome de la vena cava superior asociado al cable de marcapasos

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El síndrome de vena cava superior (SVCS) es una afectación poco común causada normalmente por neoplasias aunque, en un 20% de casos, su etiología obedece a otras causas: mediastinitis fibrosantes crónicas, tanto infecciosas como postradiación (42-70%) y trombosis venosa (6-27%). El SVCS tiene una prevalencia de 0,03% a 0,4% pero, debido al número creciente de pacientes a quienes se implantan catéteres intravenosos (marcapasos y desfibriladores), está aumentando de manera espectacular. Se presenta el caso clínico de una paciente con SVCS asociado a cable de marcapasos, cuya importancia radica en su detección y diagnóstico. Se destaca la importancia de la imagen física y se revisa las pautas diagnósticas y terapéuticas. [*Emergencias* 2009;21:151-153]

Palabras clave: Síndrome vena cava superior. Marcapasos.