CASE REPORT

Superior vena cava syndrome and syncope in an implantable cardioverter defibrillator recipient

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Abstract We describe the case of a recipient of an implantable cardioverter defibrillator with multiple syncopal episodes due to superior vena cava obstruction and electrical instability. These complications occurred in the presence of two transvenous implantable cardioverter defibrillator leads. The patient has been managed conservatively with anticoagulants and new antiarrhythmic drugs with improvement in both his clinical problems.

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KEYWORDS
syncope; defibrillator; superior vena cava syndrome; electrical storm

Introduction

Complications following the transvenous implantation of a defibrillator occur at a rate of 6.8% (2–28%). Symptomatic venous obstruction is reported in 0.5% of the cases. Symptoms usually occur in 1–15 months from implant but they may develop at any time. Venous obstruction is usually asymptomatic when a slow onset allows the development of adequate collaterals [1–3]. We report a patient with obstruction of the superior vena cava who presented multiple syncopal episodes.

Case presentation

A 50 year old man with a history of ischaemic cardiomyopathy was successfully resuscitated from cardiac arrest. Evaluation revealed ectatic coronary arteries, decreased contractility of the left ventricle (ejection fraction 30%), and a left ventricular aneurysm. Sustained monomorphic ventricular tachycardia was induced during the electrophysiological study. An implantable cardioverter defibrillator was subsequently implanted through the left cephalic vein with no complications (Jewel 7219C PCD Medtronic, Minneapolis, MN, USA, Lead: 6936 Medtronic polyurethane active fixation). The patient had few appropriate shocks for four years and was on antiarrhythmic medication, sotalol and mexiletine, during the last year. The patient underwent replacement of the exhausted generator four
years post-implantation and a new ventricular lead was implanted through the left subclavian vein due to inadequate sensing characteristics of the old one (7227CxGEM Medtronic, Lead: 6942—tined, steroid eluting tip—silicone with Sila-Cure® Medtronic). Extraction of the old lead was attempted with a lead locking device kit (Spectranetics, Colorado, USA) but was unsuccessful. Two years later the patient reported multiple syncopal episodes. These episodes were at first considered arrhythmogenic events. Careful observation during hospitalization revealed that the patient’s syncopal episodes were of two types of different origin. One group included syncope due to ventricular tachycardia/fibrillation, usually terminated by intrinsic shocks (Fig. 1). These episodes were characterized as an electrical storm because of their high frequency (> 3 episodes of ventricular tachycardia/ventricular fibrillation separated by > 5 min in less than 24 h) and were pharmaceutically managed by the combination of amiodarone, mexiletine and carvedilol. The second group of syncopal episodes had different characteristics; these were often associated with gross distension of the head and neck veins, bloating, breathlessness and blood pressure collapse and were related to changes in body position. They would easily be alleviated by lying down and elevating legs. Bedside electrocardiographic recordings during the episodes revealed sinus tachycardia at the onset and sinus bradycardia during presyncope or syncope. Actually, sporadic episodes with these characteristics had started almost immediately (a month later) after the implantation of the new electrode but had been considered as arrhythmogenic events. A CT scan and venography were performed revealing a subtotal fibroid occlusion of the superior vena cava and drainage through large and helicoid azygos (9 mm) and hemiazygos veins (Figs. 2 and 3). Pulmonary embolism was excluded. A diagnosis of superior vena cava syndrome due to subtotal occlusion of the superior vena cava was made. Although, a tilt-test was not performed, continuous heart rate and blood pressure monitoring during syncope, strongly supported this diagnosis. Thrombosis was considered the most likely responsible cause of the fibrosis. Exacerbation of symptoms over the last months was considered as an indication of an ongoing thrombotic process although this could not be proved. The helicoid anatomy of the collateral veins and the presence of two leads were considered as predisposing factors to progression of thrombosis and long-term anticoagulation therapy was initiated. Symptoms started to regress two months after its initiation.

Discussion

Lead related problems (2–22%) and inappropriate shocks are the most frequent complications in third generation implantable cardioverter defibrillators [1–4]. In asymptomatic implantable cardioverter defibrillator lead recipients significant total occlusion is encountered in 7% [2]. The superior vena cava syndrome is characterized by dyspnoea, facial and arm swelling and distension, chest wall and facial oedema which may be exacerbated by bending or lying down, syncope, presyncope, cough and cyanosis. Even in the presence of collaterals the

Figure 1 Appropriate shock for ventricular tachycardia.
venous pressure may be high enough to cause symptoms of obstruction. Venous occlusion is due to endothelial damage, fibroid reaction and blood flow perturbation which promotes thrombus formation and lead encapsulation. Occlusion occurs predominantly at locations where the lead body lies adjacent to vascular structures or where discontinuities in the lead, such as tines or the superior caval vein, contribute to the obstruction.
vena cava coil, are present. Predisposing factors for the superior vena cava syndrome are the number of leads (> 1), infections, endothelial damage during insertion, individual patient factors (i.e. deficiency of protein C, S, antithrombin III, Leiden factor) and the type of the lead [1–3]. The attempt at extraction could in itself be an additional factor predisposing to thrombosis due to exposure of tines and injury of the veins in contact with the leads. This patient, with two leads, a previous unsuccessful attempt for extraction and an extensive left ventricle aneurysm should, probably, have been considered for chronic anticoagulation, even before the onset of superior vena cava syndrome symptoms (see proximal position of the abandoned lead in Fig. 3).

A frequent problem in patients with implantable cardioverter defibrillators is electrical instability referred to as 'electrical storm'. All of the therapies delivered by implantable cardioverter defibrillators—appropriate or inappropriate—have the potential to induce ventricular and supraventricular tachyarrhythmias which is referred to as implantable cardioverter defibrillator induced proarrhythmia [4,5]. Ninety patients of 457 who received an internal defibrillator in the AVID study group [4] developed an electrical storm. Leads can induce fibroid reactions within the heart, which may result in new arrhythmogenic foci. According to Spach and Boineau [6] structural discontinuities in the heart provide a major mechanism for arrhythmias. Another possible mechanism could be the electrical interference due to noise signals between two closely implanted defibrillator leads as reported by Schulte et al. [7].

Therapeutic strategies in the presence of the superior vena cava syndrome are long-term anticoagulation, thrombolysis, stent implantation, bypass surgery, and extraction of the leads and re-implantation. We treated our patient with oral anticoagulants, although there was no hard evidence of recent thrombosis, with good results. This is in agreement with the case of Lin et al. [8] who described a case of fibrotic stenosis of the superior vena cava with sufficient collaterals and no signs of recent thrombosis; the patient received long-term anticoagulants with satisfactory results. Thrombolysis is recommended only in the case of acute onset of symptoms. When symptoms have a rapid progression, the existing venous collaterals may be inadequate and interventional therapy may be indicated. Superior vena cava stent insertion provides rapid relief of symptoms with satisfactory long-term patency rates [9]. Surgical bypass using spiral vein graft also has very good results [10]. Lead extraction has a success rate of 90%; predictors of failure are tough tissue and long existing leads [11]. Referring to the patient’s second problem which was electrical instability, treatment with antiarrhythmics is usually effective [4,5].

In conclusion, we describe the first case of an implantable cardioverter defibrillator recipient presenting with multiple syncopal episodes due to both superior vena cava obstruction and electrical instability. A conservative approach with anticoagulants and new antiarrhythmic drugs has been chosen and both clinical problems have improved. Further research is needed to document the benefit of anticoagulants in all patients with multiple leads and an unsuccessful attempt at lead extraction.

References