Facial flushing on upper limb exertion: a rare presentation of superior vena cava obstruction

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Abstract

Infection remains a significant problem for patients with cardiac-implantable electronic devices (CIEDs) but can be difficult to diagnose. We describe an unusual presentation of CIED infection in a patient with abandoned pacemaker leads. A 27-year-old male presented with facial flushing on upper but not lower limb exertion due to superior vena cava (SVC) obstruction, as well as pleuritic chest pain due to septic emboli. This was successfully treated with antibiotics and complete endovascular extraction of the pacemaker leads. Upper limb exertional facial flushing may be a useful clinical sign for the diagnosis of SVC obstruction. This case report also describes a rare presentation of CIED infection.

Introduction

Infections involving cardiac-implantable electronic devices (CIEDs) are important due to the associated high mortality [1]. We report an unusual presentation of pacemaker lead-associated endocarditis as facial flushing on upper limb exertion due to superior vena cava obstruction.

Case Report

A 27-year-old man presented to a respiratory clinic with a one-year history of episodic exertional facial flushing, scleral injection, nasal congestion, engorgement of neck and chest veins, and dyspnoea during weight training. His symptoms only occurred with vigorous upper limb exercise and did not occur during lower limb exercise such as running. Three months prior to presentation, he had pleuritic chest pains and generalized malaise.

His background was significant for Dual chamber (DDD) pacemaker implantation following chest trauma as a motorcyclist. This accident 12 years earlier caused cardiac contusion, tricuspid valve injury (treated conservatively), and complete heart block. Damage to the header was sustained in sports four years later. At this time, it was found that conduction had returned. The generator was explanted. The pacemaker leads were capped and left in-situ because of concerns regarding the risk of removing them.

Clinical examination revealed prominent veins over the anterior left chest. Facial flushing was reproduced by performing press-ups. The examination was otherwise unremarkable. He was admitted to hospital for further assessment.

C-reactive protein was 54 mg/L (normal range 0–5 mg/L). Initial computed tomography (CT) with contrast revealed multifocal areas of consolidation in both lungs, mildly enlarged bilateral hilar lymph nodes, splenomegaly, and impairment of venous drainage in the brachiocephalic veins and superior vena cava, with a filling defect suggestive of thrombus or stenosis (Fig. 1). Subsequent CT pulmonary angiogram showed no evidence of thrombus but showed that the ventricular lead had dislodged and was located in the main pulmonary artery. A transthoracic and transoesophageal echocardiogram revealed no vegetation.

Bilateral upper limb venography demonstrated moderate stenoses in the SVC and left brachiocephalic vein and
multiple collateral veins (Fig. 2). At this stage, the aetiology of SVC obstruction was unclear. Venoplasty at the time of venography was performed successfully on both stenoses. Propionibacterium acnes (P. acnes) was then identified from the admission blood cultures on day 4 of culture and on further cultures. Pacemaker lead infection was then suspected.

Treatment with intravenous ceftriaxone was initiated, and he subsequently underwent successful and uncomplicated endovascular extraction of the pacing leads. Ventricular lead damage was present. Copious stained fluid was present within the inner lumen, and lead fragments cultured P. acnes. He then received five further weeks of intravenous penicillin.

A follow-up CT chest 1 month following completion of antibiotics showed resolution of the previous areas of pulmonary consolidation (Fig. 1). At 6 months follow-up, he continued to remain well, without recurrence of previous symptoms on vigorous exertion.

**Discussion**

We report a rare presentation of SVC obstruction. This is the second case in the literature to describe SVC obstruction presenting with vigorous upper limb exercise and the absence of such symptoms with other forms of exercise [2]. In both cases, clinical symptoms and signs of SVC obstruction were reproducible in the clinic with upper limb exercise, such as press-ups. Facial flushing and venous distension produced with upper limb exercise may be a useful clinical sign in the diagnosis of SVC obstruction.

Although SVC obstruction is a known complication of chronically implanted vessels in the absence of infection, pacemaker lead infection has been implicated in the pathogenesis of SVC obstruction by inducing inflammation and subsequent fibrosis and through mechanical obstruction by thrombus [3]. We suspect that infection played a role in the development of SVC stenosis in this case. The symptoms of SVC obstruction were successfully treated by managing the infection via antibiotics and lead extraction. In the presence of CIED infection, SVC obstruction associated with implanted leads should be treated with the
extraction of all hardware. However, in the absence of infection, there are differing approaches, and often, leads may be left in-situ. The presence of symptoms of SVC obstruction occurring with vigorous upper limb exercise in a patient with a cardiac device should prompt consideration of CIED infection as a possible cause.

The most common symptoms of CIED infection include fever, chills, malaise, and local findings at the generator site, such as erythema, swelling, pain, or skin erosions [1]. Pulmonary conditions including pneumonia, recurrent chest infections, and pulmonary embolism can also be a common feature in pacemaker lead endocarditis, up to 44.7% in one case series of pacemaker lead endocarditis [4]. This case serves as a reminder that Respiratory Physicians may be referred patients with pulmonary complications from an implanted cardiac device, without the cardiac device being suspected of involvement in the presenting problem.

While the complete removal of all cardiac device hardware has been recommended as the treatment for all CIED infection [5], the management of uninfected, non-functioning, or superfluous pacing leads remains controversial. There are arguments in favour of leaving the leads in-situ, particularly if a patient has significant comorbidities, but there is concern about the longer-term risks associated with this [5]. It is thought that the risk of major complication from extraction and the difficulty of extraction increases with lead dwell time [5]. The optimal approach, especially in young patients, balancing the potential risks of extraction and the long-term risks of leaving the leads in-situ, remains a difficult decision. This case highlights the potential risks associated with leaving redundant uninfected leads in-situ and, in the absence of strong evidence-based guidance, highlights the need for individualized consideration of the risks and benefits of abandoning versus extracting redundant leads.

In conclusion, symptoms of facial flushing and venous engorgement during vigorous upper limb exercise may be a useful clinical clue to the diagnosis of SVC obstruction. In patients with abandoned pacemaker leads, symptoms of SVC obstruction and pulmonary emboli should prompt consideration of the pacemaker lead as a causative factor, warranting potential extraction.

Disclosure Statement
Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

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