CASE REPORT

An unusual case of non-malignant superior vena cava obstruction

An 81-year-old Chinese woman presented with a 1-week history of increasing facial puffiness. She had 2:1 second-degree heart block and a permanent pacemaker that had been inserted 3 years previously because of syncopal episodes. Physical examination revealed facial plethora, dilated upper trunk veins, and oedematous upper limbs suggestive of superior vena cava obstruction syndrome. This was confirmed on urgent computed tomographic scan of the thorax. There was no evidence of extrinsic compression or formation of intraluminal thrombus. The underlying aetiology was a pacemaker-induced fibrotic stricture that was successfully treated with balloon venoplasty. At 3-month follow-up, the patient remained symptom-free with normal pacemaker function.

Case report

An 81-year-old woman was admitted in March 2005 with a 1-week history of facial puffiness that had been present intermittently for the preceding few months and had not responded to diuretic therapy. There was no history of paroxysmal nocturnal dyspnoea, chest pain, chronic cough, lower limb or abdominal swelling, or frothy urine. She had a history of hypertension, cholecystectomy, chronic ischaemic heart disease, and 2:1 second-degree heart block. A permanent pacemaker had been inserted in 2002 in view of prior syncopal episodes. On examination, she had stable vital signs, facial plethora, elevated jugular venous pressure, dilated neck and upper thoracic veins, and oedematous upper limbs. Cardiorespiratory examination was otherwise unremarkable and there was no lymphadenopathy or organomegaly.

A clinical diagnosis of superior vena cava obstruction (SVCO) was made based on the triad of facial plethora, venous dilatation, and isolated upper body oedema. The possibility of malignant obstruction by tumour or lymph nodes was also high in view of the patient’s advanced age. Blood biochemistry revealed normal calcium, urate, and lactate dehydrogenase levels. An electrocardiogram revealed left ventricular hypertrophy but was otherwise unremarkable. A chest radiograph showed a normal-sized mediastinum and cardiac shadow. The lung field was clear with the pacing lead visualised in the correct position. An urgent contrast-enhanced computed tomographic (CT) scan of the thorax showed a pacemaker on the left anterior chest wall with a metallic pacing lead leading from the superior vena cava (SVC) to the cardiac chambers. A short segment of narrowing in the SVC, 4 mm in its narrowest intraluminal dimension, with dense mural calcification and eccentric wall thickening was noticed. There was no mural thrombus or evidence of extrinsic compression by a space-occupying lesion.

The patient was brought to the cardiac catheterization laboratory where an SVC venogram was performed antegradely using a pigtail catheter advanced
through the right cubital vein. Subtotal obstruction of the SVC, near its entrance to the right atrium, was visualised (Fig 1). The lesion was crossed using a 0.035” inch guidewire and SVC balloon venoplasty performed with a Jupiter Balloon (Johnson & Johnson, US) 6 mm x 20 mm at an inflation pressure of 16 atm. Satisfactory gain of blood flow from the SVC to the right atrium was observed despite a 30% residual luminal stenosis (Fig 2). No stenting was performed and the patient was prescribed warfarin. Her facial swelling subsided and she remained symptom-free at the 3-month follow-up. Pacer interrogation was unremarkable.

Discussion

Superior vena cava obstruction is a medical emergency commonly due to malignant neoplasms with extrinsic compression. The obstruction may also be a rare but serious complication that occurs following permanent pacemaker or implantable cardioverter defibrillator (ICD) implantation utilising transvenous endocardial leads. The prevalence in overseas case series varies between 0.03% and 0.4% in symptomatic patients.1,2 Most patients presented with mild facial or ipsilateral upper limb swelling instead of frank SVCO. Clinically silent pacemaker-induced stenosis is surprisingly common because of the development of an adequate collateral circulation. Severe asymptomatic SVC stenosis greater than 75% has been reported in 15% of 108 patients scheduled for ICD generator replacement by routine pre-implant venogram.3 This patient is the first reported case of pacemaker-induced SVCO in Hong Kong. Cardiac pacemaker implantation is becoming an increasingly popular procedure as its indications broaden to include conditions such as chronic heart failure and lethal ventricular arrhythmia. The incidence of pacemaker-induced SVCO can likewise be expected to increase in the near future. The accuracy of diagnosis requires a high index of clinical suspicion. An inaccurate diagnosis of acute heart failure and the subsequent prescription of intravenous frusemide can be hazardous to the patient. Given the potentially drastic clinical sequelae, physicians should routinely inform patients about the risks of SVCO when obtaining informed consent for pacemaker insertion.

The exact pathogenesis of pacemaker-induced SVCO remains unknown. Early stenosis is associated with thrombosis without a fixed stenotic lesion.4 Late stenosis is postulated to be due to fibrosis, although thrombus formation is still possible.5 The mechanical stress associated with pacemaker wires may lead to vessel wall inflammation and fibrosis, with or without thrombus formation, and ultimately, venous stenosis and occlusion. Our patient developed clinical symptoms 3 years after the implantation. Radiologi-
cases involving late development of symptomatic restenosis and pacemaker dysfunction are very rare, although one such patient has been treated successfully with a catheter-based approach.12

References

5. Lindsay HS, Chennells PM, Perrins EJ. Successful treatment by balloon venoplasty and stent insertion of obstruction of the superior vena cava by an endocardial pacemaker lead. Br Heart J 1994;71:363-5.