The incidence of dextrocardia is estimated to be 1 in ~12,000 births, while one-third of these have situs inversus. Kartagener’s syndrome is a rare condition with autosomal recessive transmission comprising situs inversus, primary ciliary dyskinesia with nasal polyposis, and bronchiectasis. Its prevalence is estimated as 1 in 130,000 of the general population. Few case reports of catheter ablation of supraventricular tachyarrhythmias in patients with dextrocardia and situs inversus published. This report is the first to provide a three-dimensional electroanatomical map of a patient with the Kartagener’s syndrome and atrioventricular nodal reentrant tachycardia. Tagging the atrioventricular node and the slow pathway on the three-dimensional electroanatomical map was useful to avoid atrioventricular block in the presence of unusual anatomy. Likewise, novel technologies for remote robotic catheter navigation, such as magnetic navigation, may be helpful in such cases.

Left atrial isomerism with azygous continuation and interruption of the inferior vena cava may be observed unexpectedly during insertion of electrophysiology catheters. Isomerism is present in 1–4% of patients with congenital heart defects with a large heterogeneity at clinical presentation depending on associated cardiac malformations. Intracardiac involvement in left atrial isomerism is often less severe than in right atrial isomerism. The presence of interrupted suprarenal portion of the inferior vena cava with azygous continuation strongly suggests left atrial isomerism, which is frequently associated with two or more spleens (polysplenia syndrome) as it was the case in our patient.

During a standard catheter ablation procedure, the catheter is targeted to the ablation region based on typical local electrogram characteristics and anatomical landmarks. As indications for catheter ablation expand, the complexity of the anatomical substrates in patients with congenital anomalies may pose a challenge to the electrophysiologist. Our cases illustrate the role of imaging in patients with unusual presentation of the anatomy of the cardiac chambers and major blood vessels. The identification of the accurate anatomy using imaging modalities such as computerized tomography and magnetic resonance imaging and three-dimensional image reconstruction using mapping systems may be useful not only to help the electrophysiologist enabling a safe and successful catheter ablation procedure, but also to understand the complex anatomical structures and to guide for optimal catheter access.

References


Massive thrombi on an implantable cardioverter-defibrillator lead in a patient with the antiphospholipid syndrome

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**CASE REPORT**

A patient with the antiphospholipid syndrome, who was suboptimally anticoagulated, presented with two large thrombi attached to her implantable cardioverter-defibrillator lead. Anticoagulation was unsuccessful so, in view of the risk from embolization, the system was removed surgically and an epicardial system implanted to reduce the risk of future device-related thrombosis.

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A 48-year-old woman with a previous pulmonary embolism, two stillbirths, and who was persistently positive for lupus anticoagulant antibody had a myocardial infarction in 2003 in the context of angiographically normal coronary arteries and a year after this a single-chamber implantable cardioverter-defibrillator was implanted for primary prevention of sudden cardiac death. She failed to achieve a target international normalized ratio (INR) of 2–3 with oral vitamin K antagonists and consequently for the 7 months before presentation had taken treatment-dose subcutaneous enoxaparin once daily. She was admitted with chest pain and breathlessness, although the ventilation-perfusion lung scan had a low likelihood of pulmonary embolism. However, on echocardiography there were two large masses, 3.1 and 3.4 cm in diameter, attached to the defibrillator lead (Figure 1). These were felt likely to be thrombi (confirmed on subsequent histology). A 2-week trial of in-hospital warfarin therapy (to circumvent possible compliance issues) with intravenous heparin to bridge anticoagulation and a target INR of 3–4 was attempted but produced no change in the echocardiographic appearance. With conservative management not having succeeded and in view of the size of the thrombi with the consequent risk posed were they to embolize, as well as the possibility from the presentation that embolization had already occurred to a lesser degree, the pacing system was removed surgically. To reduce the risk of future device-related thrombosis, an epicardial system was implanted at the time of surgery. The patient was discharged after an uncomplicated post-operative stay on warfarin with a target INR of 3–4.

Antiphospholipid syndrome is characterized by thrombosis (which can be arterial or venous) or morbidity in pregnancy in the presence of a persisting antiphospholipid antibody. Antiphospholipid antibodies are detected by lupus anticoagulant or anticardiolipin or anti-beta2 glycoprotein 1 antibody testing: only one test needs to be positive but in order to ensure that this is not transient or a false positive the test needs to be repeated after 8–12 weeks. In those with previous venous thrombosis, anticoagulation to an INR of 2–3 is recommended, while in those with arterial thrombosis, or recurrent venous events, an INR above three has been suggested. The most common cardiac manifestation of the syndrome is valvular disease, but cases of intracardiac thrombosis have been reported for all cardiac chambers.

Pacing lead-related thrombosis is a well-recognized phenomenon and the management options are anticoagulation to allow the thrombus to dissipate or surgical extraction. If the thrombus is asymptomatic, at low risk of embolism and not so large that the consequences of embolism would be serious, then anticoagulation is a reasonable option with reported dissolution of thrombus at 4 weeks in one case using warfarin. If the thrombus is symptomatic or at high risk of embolizing or from the consequences of embolization, as in our case, then surgical intervention is warranted. In an isolated case in which anticoagulation was unsuccessful but surgery was deemed too high risk, prolonged low-dose tissue-type plasminogen activator infusion was successful.

The case highlights the need for vigilance in thrombophilic patients with respect to device-related thrombosis. In such patients known to be poorly compliant with oral treatment, initial implantation with an epicardial or subcutaneous rather than an endocardial system should be considered.

Conflicts of interest: none declared.

References