Metastatic Merkel cell carcinoma of the heart

Louis W. Wang1,2*, Bruce D. Walker1,2, Abdullah Omar3,4, Andre E. Tay1, and Rajesh N. Subbiah1,2,5

1Department of Cardiology, St Vincent’s Hospital, Sydney, NSW, Australia; 2St Vincent’s Clinical School, University of New South Wales, Sydney, NSW, Australia; 3Department of Vascular Medicine, St Vincent’s Hospital, Sydney, NSW, Australia; 4Faculty of Health, University of Technology, Sydney, NSW, Australia; and 5Cardiac Electrophysiology and Biophysics Program, Victor Chang Cardiac Research Institute, Sydney, NSW, Australia

* Corresponding author. Tel: +61 283821111, Fax: +61 283822359, Email: louis.wang@unsw.edu.au

A 76-year-old man with ischaemic cardiomyopathy and a cardiac resynchronization therapy defibrillator presented after a witnessed collapse. The patient denied any cardiorespiratory symptoms, but reported recent lethargy and weight loss. Two years earlier, he was diagnosed with Merkel cell carcinoma of the left neck, successfully treated with modified radical neck dissection and adjuvant radiotherapy. Cardiac resynchronization therapy defibrillator interrogation revealed 14 episodes of non-sustained ventricular tachycardia in the 6 months prior to presentation. Although atrial and right ventricular pacing thresholds remained stable since implantation, left ventricular pacing lead threshold had increased (from 2.0 to 3.2 V; 0.4 ms pulse width). Chest X-ray revealed stable lead positions. Transthoracic and transoesophageal echocardiography (Panels A and B, Supplementary material online, Videos S1–3) demonstrated a large mass in the coronary sinus, encasing the left ventricular pacing lead, which was confirmed on chest computed tomography (Panels C–E). Whole body positron emission tomography–computed tomography showed an extensive intra-cardiac focus of 18-fluorodeoxyglucose avidity and a second focus in a lymph node adjacent to the pancreas (Panels F and G). Endoscopic ultrasound with fine needle aspiration of this lymph node was performed. Immunohistochemistry confirmed neuroendocrine carcinoma of Merkel cell origin. At completion of palliative carboplatin and etoposide chemotherapy, left ventricular pacing lead threshold returned to baseline. Merkel cell carcinoma is a rare, aggressive neuroendocrine tumour. This case describes an unusual electrophysiological manifestation of metastatic Merkel cell carcinoma of the heart. The increase in ventricular tachycardia may have been related to direct tumour infiltration, while the changes in left ventricular pacing threshold resulted from tumour disruption of the normal pacing lead–myocyte electrical interface.

All authors contributed to clinical care and writing of the manuscript.

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