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Combined Mechanism of Refractory Shock in Primary Cardiac Lymphoma: A Rare Dilemma

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Primary cardiac lymphoma is very rare, and usually manifests after the fifth decade of life. The lack of typical manifestations makes it difficult to diagnose at an early stage that can be discovered only by echocardiography. The location of the tumour often results in cardiac compromise, which prevents the delivery of potentially curative therapies. Clinical presentations may depend on flow obstruction, infiltration of adjacent tissues, tumour embolisation, and atrioventricular (AV) disturbances. We report a rare case of primary cardiac lymphoma that presented with clinical signs of shock from two distinct mechanisms. The first mechanism was intermittent complete AV block that was caused by disruption of the electrical conduction system from tumour infiltration in addition to direct mechanical compression of the atrioventricular node by the tumour. The second mechanism, subtotal RV inflow obstruction from the bulky mass contributed to compromising venous return, which played a major role of refractory shock in this case.

Keywords Complete atrioventricular block • Shock • Primary cardiac lymphoma • Echocardiography

Introduction

Primary cardiac lymphoma is very rare, and usually manifests after the fifth decade of life [1]. It has been reported in only 1% of primary cardiac tumours and 0.5% of extranodal lymphoma [2]. The lack of typical manifestation makes it difficult to diagnose in the early stage [3] that can be initially discovered only by echocardiography. The delay in diagnosis results in the poor prognosis despite chemotherapy [4]. Clinical presentations may depend on flow obstruction [5], infiltration of adjacent tissues, tumour embolisation, and atrioventricular disturbances [6,7] where the right atrium (RA) is the commonest site involved. The location of the tumour often results in cardiac compromise, which prevents the delivery of potentially curative therapies. In some rare cases, the patient can present with solitary restrictive cardiomyopathy from extensive infiltration of atypical lymphocytes without demonstrable intracardiac mass [8]. We report a case of primary cardiac lymphoma presented with clinical signs of shock from two distinct mechanisms.

Clinical Presentation

A 70 year-old male presented with acute onset of lightheadedness. He reported two-month progressive anorexia and weight loss. He had a history of status post right

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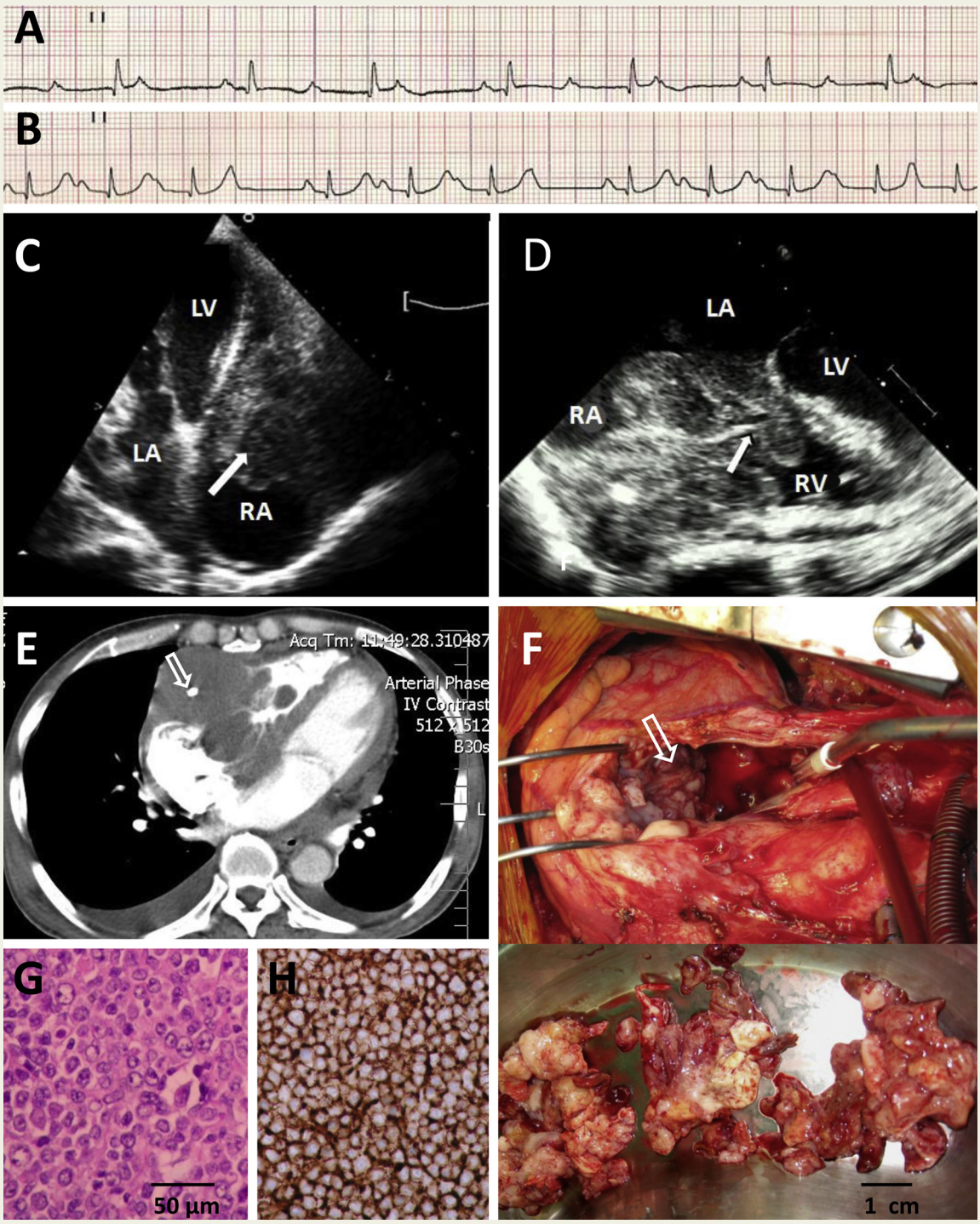

Figure 1 (A) Initial electrocardiogram (ECG) demonstrating complete AV block with junctional escape of 52 beats/min. (B) ECG showing the downgrading of complete AV block to Mobitz-I second degree AV block of 92 beats/min. (C) Transthoracic echocardiography unveiling a huge mass (arrow) occupying the right atrium which extending into the right ventricle causing a near total obstruction of tricuspid inflow. LA indicates left atrium; RA, right atrium; LV, left ventricle and RV, right ventricle. (D) Transoesophageal echocardiography revealing a large echogenic mass involving the interatrial septum and protruding in and out between the tricuspid valve (arrow). (E) Computerised tomography demonstrating a non-enhanced hypodensity mass infiltrating from epicardium through endocardium of the right-sided cardiac chamber with marked thickening of the interatrial septum (arrow shows right coronary stent). (F) Intraoperative finding (above) showing the impacted tumour (arrow) in right atrium from which the vast majority of the friable tumour was excised (below). (G) Histological examination demonstrating diffuse proliferation of abnormal large lymphoid cells (hematoxylin and eosin stain). (H) Positive immunohistochemical staining of CD20 (B-cell marker), consistent with B-cell lymphoma.