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Letter to the Editor

Coronary vessel floating sign and vasospastic angina in a patient with cardiac lymphoma

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A 59-year-old immunocompetent man was referred to our emergency room with syncope. The patient's Glasgow Coma Scale was E4V5M6, and neurological examination revealed no palsy. He reported no history of trauma or prescribed medication use, and his vital signs were as follows: body temperature, 37.3 °C; blood pressure, 104/74 mm Hg, pulse rate, 30 beats per minute (bpm) with irregular rhythm; and oxygen saturation in room air, 100%. Physical examination revealed distention of the jugular vein in the sitting position. On auscultation, his lung sounds were normal, and the pericardial friction rub was not clearly heard. Lower leg edema was not evident. Chest X-ray revealed cardiomegaly without pulmonary congestion. Electrocardiography (ECG; 12-lead) showed atrial fibrillation with bradycardia and ST elevation in the inferior leads. Soon after admission, atrial fibrillation spontaneously restored to sinus rhythm, and ST elevation also improved (Fig. 1A and B). Emergency coronary angiography revealed no significant stenosis (Fig. 2A and B; see Supplementary Videos 1 and 2). We suspected vasospastic angina (VSA) because of ST elevation without coronary artery stenosis. Laboratory findings were as follows: white blood cell count, 11,400 mm^3; C-reactive protein, 3.9 mg/dL; procalcitonin, 0.06 ng/mL; creatine phosphokinase, 165 IU/L; troponin T, negative; and brain natriuretic peptide, 15.7 pg/mL. Thyroid function was normal and antinuclear antibody was negative. The lactate dehydrogenase (LDH) level was slightly elevated (422 IU/L), the serum soluble interleukin-2 receptor (sIL2R) level was within normal limits (445 U/mL). Paradoxical pulse became overt in sinus rhythm. Transthoracic echocardiography revealed circumferential pericardial fluid retention (Fig. 3 and Supplementary Video 3). Pericardiocentesis yielded 1 L of bloody pericardial fluid containing an extremely high LDH (7752 IU/L) level. Cytological examination of pericardiocentesis fluid revealed numerous mid- to large-sized atypical lymphoid cells with prominent nucleoli. Subsequent ECG-gated, contrast-enhanced multidetector row computed tomography (MDCT) revealed a mass located around the atrioventricular (AV) groove and encasing the right coronary artery (RCA) without arterial invasion or compression (vessel floating sign; Fig. 4A–D). The tumor extended to the inferoposterior wall of the left ventricle (LV) along the RCA (Fig. 4E and F). Abnormal accumulation was observed at the same site on a gallium 67 scan. Immunohistochemical analysis of the cell block section of the effusion showed that the lymphoid cells were positive for CD20, with a high MIB-1 index, and negative for CD3. A final diagnosis of B-cell lymphoma was made (Fig. 5A and B). Complete remission of the tumor was achieved after 6 cycles of rituximab, cyclophosphamide, hydroxydaunorubicin, oncovin, and prednisone (R-CHOP) therapy (Fig. 6A–F). An acetylcholine (Ach) provocation test by injection of Ach into the coronary artery after tumor remission due to R-CHOP therapy did not induce coronary spasm (Fig. 2C and Video 4).

Primary cardiac lymphoma (PCL) is a rare entity with variable clinical presentations. Although PCL is more common in immunocompromised patients, it is exceedingly rare in the immunocompetent population. The presenting symptoms are often nonspecific because they may vary according to the heart sites involved by PCL. The most frequent cardiac manifestations are pericardial effusion, heart failure, and AV block. PCL sometimes follows a fulminant course and is associated with a high mortality rate because of pulmonary embolism, arrhythmia, and heart failure [1]. Patients with malignant lymphoma occasionally experience ischemic heart disease (IHD) due to complications of lymphoma therapies. A causative link between radiation therapy to the mediastinum and coronary artery disease is well recognized in patients with Hodgkin lymphoma [2]. Also, Vinca alkaloids and granulocyte colony stimulation factor are suggested to be associated with IHD [3,4]. However IHD-related lymphoma itself is very rare. Lymphoma can reportedly cause IHD due to direct

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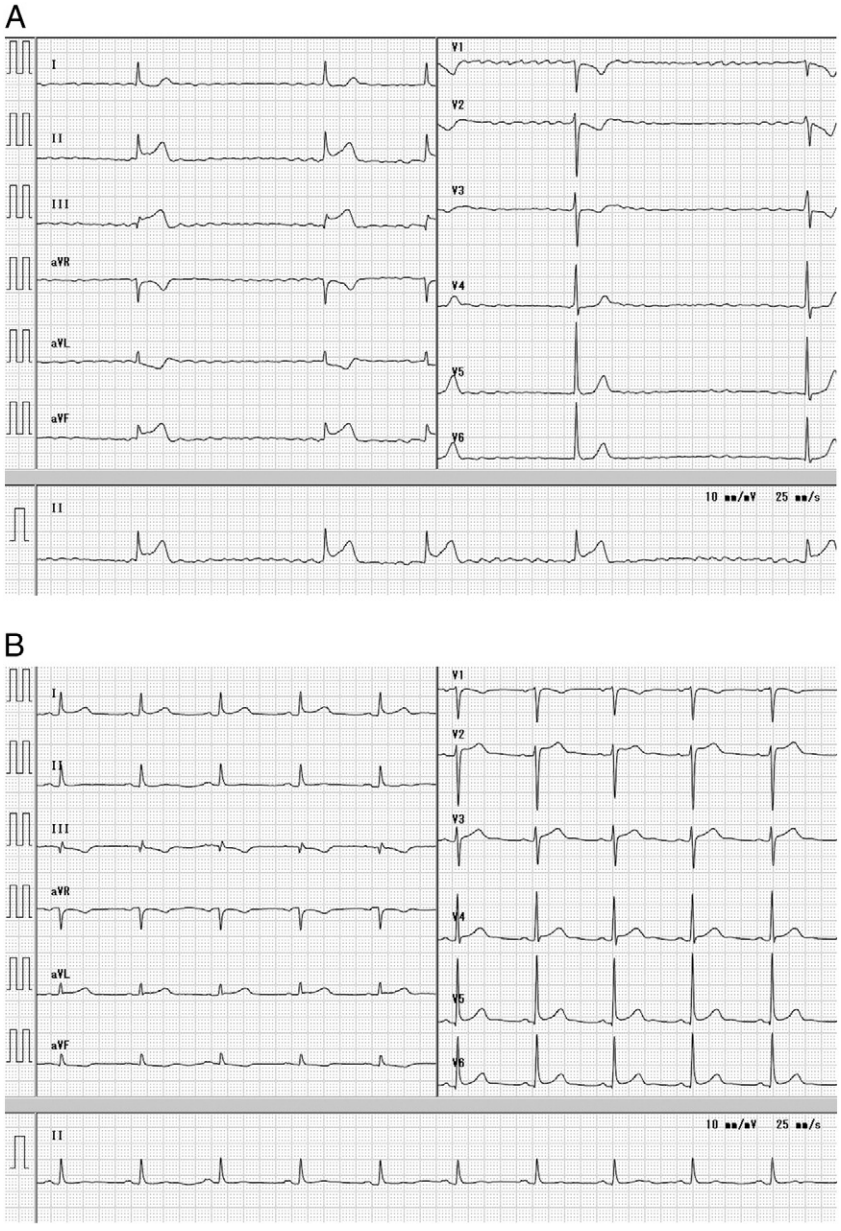

Fig. 1. Twelve-lead electrocardiography reveals atrial fibrillation with bradycardia; ST elevation in inferior leads II, III, and aVf; and reciprocal ST depression in leads V1–3 (A). Atrial fibrillation spontaneously restored to sinus rhythm, and ST elevation also improved (B).

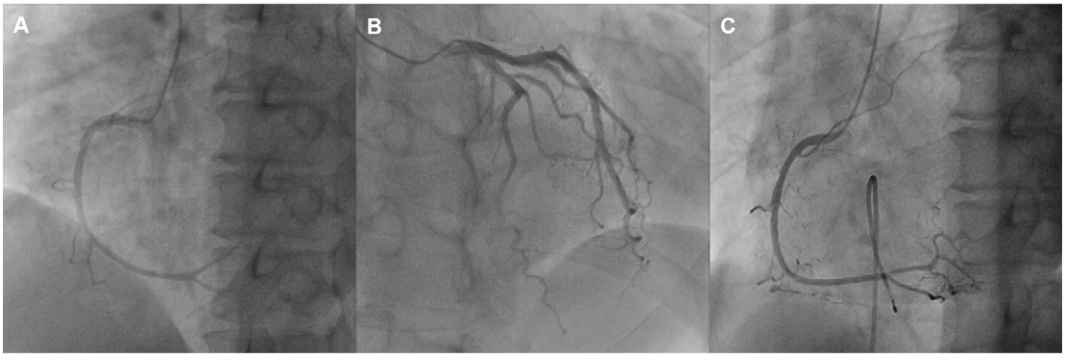

Fig. 2. Emergency coronary angiography shows no significant stenosis in the right coronary artery (RCA) in the right anterior oblique view (A and Supplementary Video 1) and left coronary artery in the left anterior oblique view with cranial angulation (B and Supplementary Video 2). Injection of 50 μg of acetylcholine into the RCA in an acetylcholine provocation test performed after tumor remission does not induce coronary artery spasm (C and Supplementary Video 4).