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Images in Cardiovascular Medicine

A Long Journey to the Truth: Primary Cardiac Lymphoma with Various Arrhythmias from Ventricular Tachycardia to Atrial Flutter

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On May 11, 2010, a 43-year-old man visited our hospital with palpitations and dizziness for one year. Initial electrocardiogram (ECG) showed first and second degree atrioventricular block (**Figure 1A and B**). At first, transthoracic echocardiogram (TTE) was normal (**Figure 2A**). Over a 7-year period, ECG findings gradually proceeded and various arrhythmias developed including ventricular tachycardia (VT) (**Figure 1C-F**). TTE showed diffuse progressive ventricular and atrial wall thickening (**Figure 2B and C**). Finally, TTE expressed a huge mass in right atrial cavity (**Figure 2D**). A total of 3 transvenous endomyocardial biopsies were performed and all revealed no abnormal findings. Chest computed tomography (CT) showed huge cardiac masses and anterior mediastinal lymph node enlargement (**Figure 3**). He was initially considered as hypertrophic cardiomyopathy and then cardiac sarcoidosis. On April 5, 2017, anterior mediastinal lymph node excision biopsy was conducted and the pathology was malignant large B cell lymphoma (**Figure 4**). He was finally diagnosed with primary cardiac lymphoma (PCL). Rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone chemotherapy was performed. On positron emission tomography-CT after 9 months of chemotherapy, ^{18}F-fluorodeoxyglucose uptake significantly decreased (**Figure 5**) and VT no longer occurred. TTE findings became almost normal (**Figure 2E**).

PCL is a rare disease composing 1.3% of primary cardiac tumors.[1] PCL has no pathognomonic presentation, manifested by location. Intramural tumors can present with fatal arrhythmia such as VT or conduction defects[2-4] at the beginning by invading the conduction system even before the structural lesion is detected. Continuous questions of VT mechanism and constant biopsies finally reached the proper diagnosis. And active treatment of intractable VT with amiodarone, implantable cardiac defibrillator and radiofrequency catheter ablation prevented death of the patient.

The informed consent that allows the publication of clinical data was obtained from the patient.

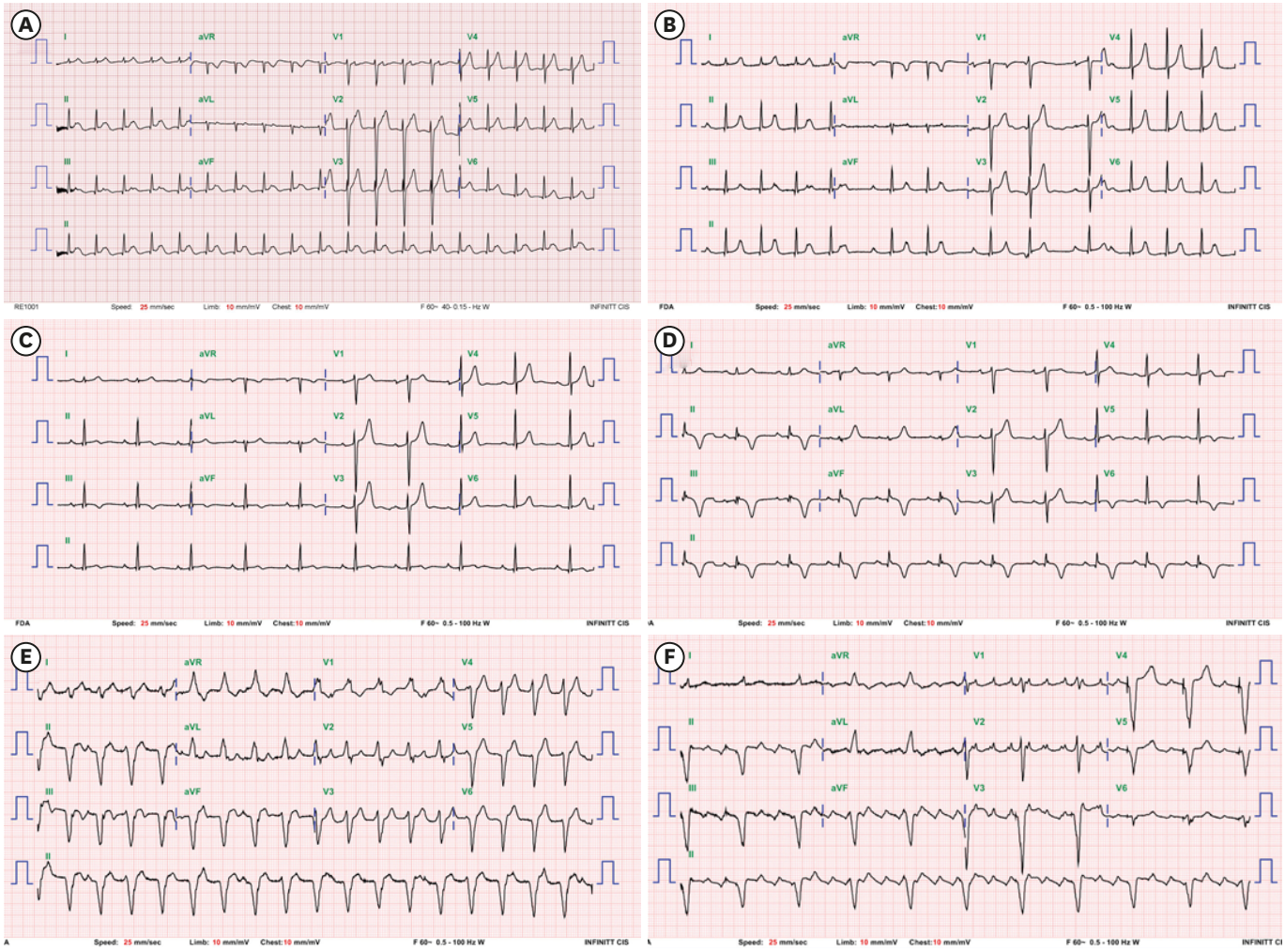

Figure 1. Twelve-lead ECGs. (A) ECG at the time of the first hospital visit showed 1st degree AV block (PR interval 360 ms). (B) Mobitz type I second-degree AV block was also recorded. (C) In the 3rd year after the first visit, T inversion began to be observed. (D) In the 4th year after the first visit, deep T-inversion proceeded in inferior leads and lateral precordial leads. (E) In the 5th year after the first visit, intractable ventricular tachycardia developed. (F) In the 7th year, after steroid pulse therapy for sarcoidosis, a typical atrial flutter occurred.
AV = atrioventricular; ECG = electrocardiogram.

Conflict of Interest

The authors have no financial conflicts of interest.

Author Contributions

Supervision: Choi Y, Oh YS, Kim SH; Writing - original draft: Kim S; Writing - review & editing: Kim YR.

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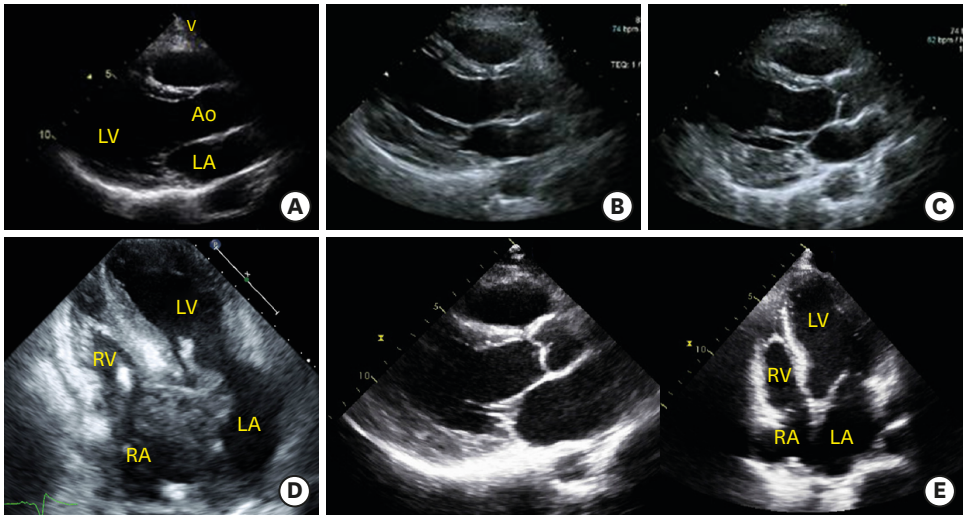

Figure 2. Images of transthoracic echocardiography. (A) Transthoracic echocardiogram at the time of the first hospital visit showed normal findings. (B) In the 4th year after the first visit, it indicated hypertrophied posterior wall thickness with hyperechoic myocardium. (C) In the 5th year after the first visit, unusual pattern of LV inferolateral wall hypertrophy was shown. (D) In the 7th year after the first visit, more progression of asymmetric LV hypertrophy and a huge mass in RA cavity with near-total obstruction of flow across tricuspid valve were observed. (E) Nine months after chemotherapy, the huge mass involving interatrial septum disappeared and LV wall thickness was nearly normalized.
Ao = aorta; LA = left atrial; LV = left ventricular; RA = right atrial; RV = right ventricular.

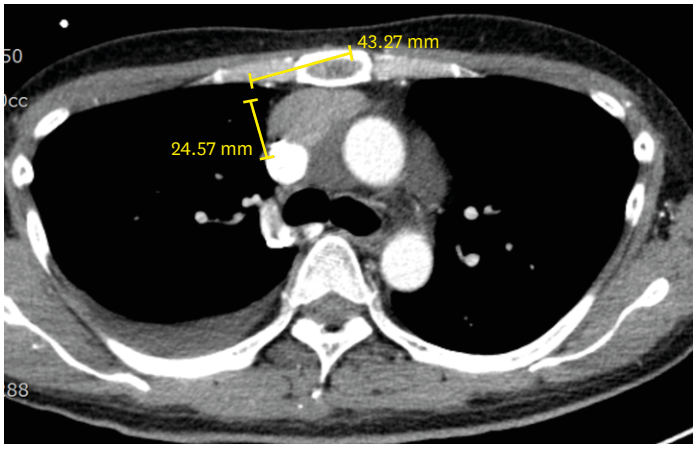

Figure 3. Chest computed tomography in the 7th year after the first hospital visit revealed anterior mediastinal lymph node enlargement.

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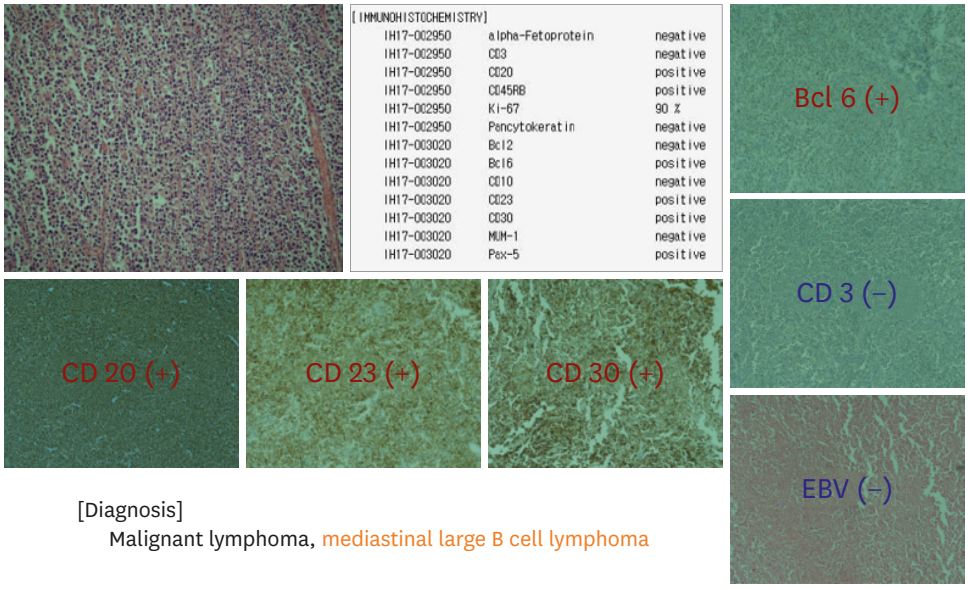

Figure 4. Anterior mediastinal lymph node excision biopsy showed a malignant large B cell lymphoma.
Bcl = B-cell lymphoma; CD = cluster of differentiation; EBV = Epstein-Barr virus.

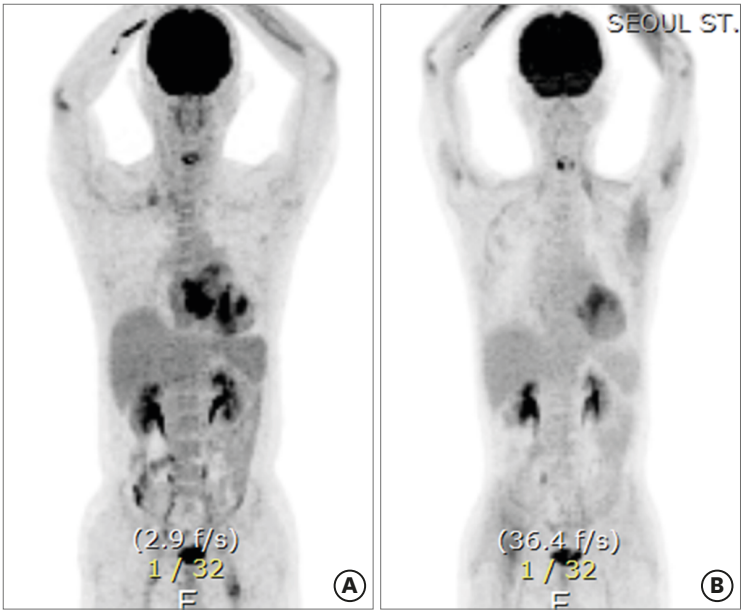

Figure 5. Images of positron emission tomography-computed tomography. (A) In the 7th year after the first visit, intense focal FDG uptakes were observed in the LA and LV wall. (B) Six months after starting rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone chemotherapy, the FDG uptake in the LV wall decreased significantly.
FDG = fluoro-deolyglucose; LA = left atrial; LV = left ventricular.

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