Chest Pain, Shortness of Breath, and Palpitations Unmask an Unexpected Diagnosis

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A 48-year-old white man was admitted to our hospital because of a sustained wide-complex tachycardia (160 bpm). He presented with chest discomfort, dyspnea, palpitations, and dizziness. Because medical cardioversion with intravenous amiodarone failed, intravenous ajmaline was administered, which successfully resulted in conversion to sinus rhythm. Physical examination and laboratory results revealed no significant abnormalities. The patient reported no discomfort until the morning of admission, and there was no history of disease except for tuberculosis, successfully treated 20 years earlier. Although serial cardiac troponin T analysis was negative, the patient complained of ongoing angina. Cardiac catheterization showed subtotal stenosis of the right coronary artery (Figure 1A), which was successfully treated by percutaneous coronary intervention and placement of a bare-metal stent (Figure 1B). Notably, the coronary arteries showed no high-grade lesions in the other main coronary vessels. Echocardiography revealed an echodense mass at the right ventricular lateral wall, the interatrial and interventricular septum, and the left atrial wall. A magnetic resonance imaging scan (Figures 2A and 2B; online-only Data Supplement Movies I–IV) calculated a tumor volume of \( \approx 130 \text{ mL} \) and showed mild pericardial and pleural effusions. Left and right ventricular ejection fractions were slightly impaired (50% and 49%, respectively), and a right ventricular end-diastolic volume of 135 mL was calculated. Because contrast-enhanced computed tomographic coronary angiography showed the right coronary artery coursing through the tumor mass (Figure 2C), lumen narrowing was anticipated to be caused by tumor compression. Histopathology of several biopsy samples obtained by ministernotomy provided evidence of follicular lymphoma of World Health Organization grade II (Figures 3A through 3D). In follicular lymphoma, secondary involvement of the heart in systemic disease occurs 20 to 40 times more often than primary cardiac origin. However, staging by neck, thoracic, and abdominal computed tomography and positron emission tomography scans and bone marrow biopsy ruled out systemic disease, thus establishing the diagnosis of primary cardiac lymphoma, stage IE. Notably, an HIV test was negative.

Generally, primary cardiac lymphomas are quite rare (\( \approx 200 \) cases reported in the literature) and in most cases are...
diagnosed at autopsy. They usually progress quickly and first involve the right side of the heart and pericardium. The prognosis is poor and often is worsened by delay in diagnosis. According to the literature, the majority of primary cardiac lymphoma patients have various nonspecific symptoms, such as dyspnea, edema, arrhythmia, cardiac tamponade, and congestive heart failure. To the best of our knowledge, initial manifestation with acute coronary syndrome has not been reported to date.

Current recommendations suggest treatment of indolent primary cardiac lymphoma–like localized lymphomas that arise on other primary sites. Accordingly, radiation therapy is the first choice for treatment of follicular lymphoma stage I or II. However, the extended cardiac involvement in the present case gave rise to concerns about toxicity and about the immediate effect of radiation-induced tumor regression. Hence, we decided to treat the patient with the anti-CD20 antibody–containing immunochemotherapy regimen R-COP (rituximab, cyclophosphamide, vincristine, and prednisone) for 6 cycles, to avoid an acute and calculable risk to the patient. Doxorubicin was not used because of the well-known cardiotoxicity of anthracyclines. During the onset of chemotherapy, the patient displayed recurrent wide-complex tachycardias (Figure 4A) whenever continuous intravenous

Figure 2. A, MRI demonstrates an isointense mass on steady-state free precession images of the 4-chamber view that infiltrates the interventricular and interatrial septum, right ventricle, and left atrium (arrows). B, The intracardiac mass was hyperintense on fat-suppressed short-axis T2-weighted images. C, Contrast-enhanced computed tomographic coronary angiography shows the right coronary artery (RCA) coursing through the tumor mass located within the free wall of the right ventricle. Note the localization of the coronary stent at segment 2 of the RCA, indicated by arrow. RA indicates right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; and Ao, aorta.

Figure 3. Histology shows infiltration of myocardium and epicardial fat by nodular and diffusely growing atypical B cells. A, Hematoxylin-and-eosin stain (original magnification ×100). B, Giemsa stain (×400). In addition to numerous centrocytes, classic centroblasts are recognized. C, CD20 immunostain (×200). D, Bcl-6 immunostain (×400).
Ajmaline administration was paused but developed Mobitz I block (Figure 4B) under infusion of the drug. A single-chamber cardioverter defibrillator was implanted to prevent sudden death, and ajmaline was administered orally. Ongoing chemotherapy could then be continued, and complications ceased. Staging after 3 cycles revealed no further tumor enlargement, although the lesion did not diminish. Against our recommendation, the patient discontinued chemotherapy after 3 cycles, mainly because of his improved medical status. The 6-month follow-up revealed stable disease. At

Figure 4. ECG documentation of arrhythmic complications. A, Wide-complex tachycardia (120 bpm). B, Second-degree Mobitz I AV block (44 bpm).

Figure 5. Comparison of the 2 computed tomography scans: A, at baseline (before chemotherapy); B, 2 years after administration of last cycle of chemotherapy, showing significant tumor regression, which indicates remission of the disease.
2 years, the patient presented without heart failure or any ischemic or arrhythmic complications. The follow-up computed tomography scan showed significant tumor regression compared with baseline acquisitions (Figures 5A and 5B).

The present case underlines the importance of early diagnosis by multilevel imaging and prompt oncological intervention in the management of primary cardiac lymphoma. Furthermore, continuous cardiological monitoring is indispensable to avoid life-threatening complications that arise from changes in tumor size.

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Disclosures
None.

References
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