

Primary Cardiac Lymphoma Presenting with Atrial Fibrillation

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Atrial fibrillation is a common condition affecting approximately 2.6 million people in the United States. Disorders commonly associated with AF include structural heart disease (i.e., hypertrophy and valvular disease), ischemic heart disease, and hyperthyroidism [1]. A rare cause of cardiac arrhythmias in general and AF in particular are cardiac tumors [2].

Most cardiac tumors are metastatic; primary cardiac tumors are rare (prevalence of 0.001–0.028%), most being benign myxomas. Approximately 25% of primary cardiac tumors are malignant, 75% of which are sarcomas and approximately 6% lymphomas [3]. We present a case of primary cardiac lymphoma presenting with AF and dyspnea.

PATIENT DESCRIPTION

A 66 year old woman with a known history of hypertension was admitted to an internal medicine ward complaining of palpitations and shortness of breath. The patient had been discharged from the same department 7 days earlier after a brief hospitalization following typical atrial flutter that converted to sinus rhythm following administration of verapamil. At the time of the first hospitalization palpitations had

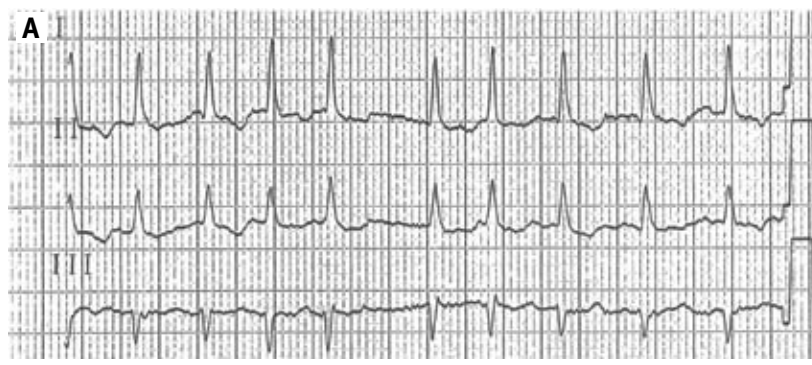
been present for approximately 3 weeks and were attributed to considerable life stress. Although the patient complained of dyspnea there was no hypoxia or signs of respiratory distress on physical examination. The patient was discharged on verapamil, coumadin and aspirin with a recommendation for an ambulatory echocardiogram and repeated lactate dehydrogenase measurement in light of an elevation to 759 U/L.

At admission the patient had a pulse of 105, blood pressure 136/89 mmHg, respirations 14. She was in a generally good condition, without cyanosis. There was no elevation of jugular venous pressure, a few diffuse crackles were heard over both lung fields, and heart sounds were irregular with no associated murmurs. Electrocardiography demonstrated atrial fibrillation [Figure A] with no signs of ischemia. Chest X-ray did not demonstrate pulmonary congestion; however, the cardiac silhouette was enlarged. Pertinent laboratory values included: calcium 11.9 mg/dl, parathyroid hor-

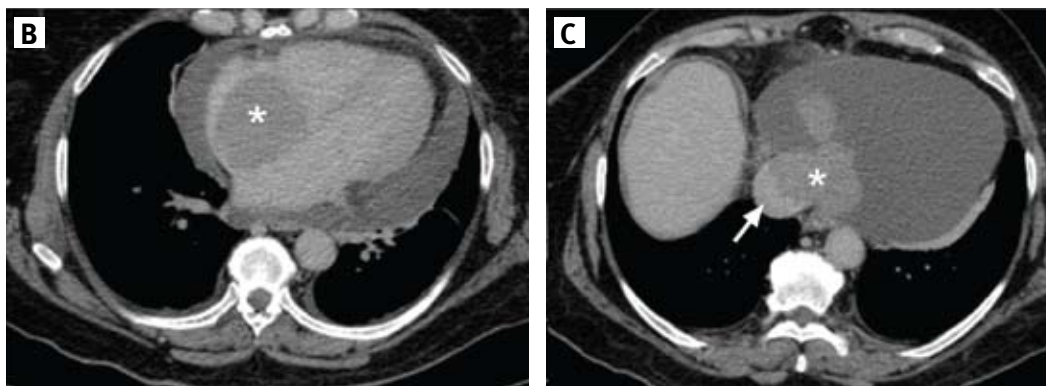
mone 47.3 pg/ml, alkaline phosphatase 169 U/L, aspartate aminotransferase 76 U/L, alanine aminotransferase 123 U/L, lactate dehydrogenase 731 U/L, thyroid-stimulating hormone 2.9, troponin T-B < 0.01 ng/ml and international normalized ratio 2.97. Arterial blood gases were as follows: pH 7.51, PO₂ 64, PCO₂ 24.3 and HCO₃ 19.6.

Due to the combination of dyspnea and hypoxia, further tests were performed mainly to rule out pulmonary embolism. Lower extremity duplex showed no evidence of deep vein thrombosis. Echocardiography demonstrated an echogenic mass in the right atrium measuring 52 x 32 mm; an additional mass external to the right atrium was observed (26 x 39 mm) that contained blood flow, reflecting invasion of the inferior vena cava. In addition, a large pericardial effusion was demonstrated. Computed tomography angiography showed a space-occupying lesion with lobular borders in the right atrium extending into the coronal sinus and the inferior vena cava (8.4 x 6.6 x 9.0

[A] ECG leads I-III during atrial fibrillation



AF = atrial fibrillation



[B,C] CT angiography showing mass in the right atrium (marked with an asterisk) **[B]**, and extending into the inferior vena cava (arrow) **[C]**.

cm) [Figures B & C]. Coumadin treatment was terminated to reduce the risk of pericardial bleeding and tamponade formation, and the patient was transferred to a tertiary hospital that had a heart and chest surgery department.

Pericardiocentesis was performed with drainage of approximately 1000 ml of blood-stained fluid. Cytology of the fluid revealed intermediate sized cells with irregular nuclear membranes and coarse chromatin; a few small lymphoid cells were also seen. Immunocytostains were positive for leukocyte common antigen and CK22 (cytokeratins 22) but negative for CD20, CD3 and carcinoembryonic antigen; these measures were not sufficient for diagnosis. Blood drawn for tumor markers was positive for CA-125 (107.66) and negative for CA-19-9 and CEA. Transthoracic and transesophageal echocardiograms confirmed the presence of a giant tumor in the right atrium extending into and partially obstructing the inferior vena cava and coronary sinus, with a significant right to left shunt through a patent foramen ovale; there was mild tricuspid regurgitation 2.5 m/sec without evidence of pulmonary hypertension. A biopsy was deemed necessary in order to reach a final diagnosis, although a percutaneous approach was excluded due to significant risk for systemic embolization through the patent foramen ovale. An open biopsy was performed through a sternotomy and samples were sent for fro-

zen sections. The intraoperative pathology report suggested lymphoma. Therefore, the operation was concluded without an attempt at removal or debulking. The final pathology report confirmed a diagnosis of diffuse large B cell lymphoma with the following immunostaining results: LCA+, CD20+, CD30-, ALK1- (anaplastic lymphoma kinase 1), CEA-, TTF1- (thyroid transcription factor-1), CK22-, CD43-, CD3 +/- . The postoperative course was complicated by significant respiratory distress with blood oxygen desaturation. Corticosteroids and allopurinol were administered following consultation with the hematologist, as were nitric oxide and milrinone to treat an increase in the right to left shunt. These measures led to clinical improvement.

The patient was transferred to hematology care for chemotherapy that included cyclophosphamide followed by a full course of CHOP (cyclophosphamide, adriamycin, vincristine and prednisone). The post-chemotherapy period was complicated by hemodynamic and respiratory aggravation requiring mechanical ventilation and vasopressors; however, her condition improved and she continued with chemotherapy. Follow-up echocardiography demonstrated a reduction in the size of the tumor.

COMMENT

Primary cardiac lymphoma is rare, as exemplified by the unusual case described here, which presented with arrhythmia as

the primary presenting symptom. Due to the low incidence of cardiac tumors in general, and cardiac lymphoma specifically, a high index of suspicion is required if lymphoma is to be suspected in a patient presenting with AF. The American Heart Association/American College of Cardiology guidelines for the evaluation of patients with AF [4] require an echocardiography to assess left heart dimensions and function and to exclude occult valvular or pericardial disease and hypertrophic cardiomyopathy. However, in daily practice this is often deferred to the ambulatory setting; thus a patient with AF may be discharged from the emergency department without any cardiac imaging that might hint at a tumorogenic etiology. In the presence of specific alarming characteristics such as cardiomegaly on chest X-ray or respiratory distress, earlier echocardiography should be considered.

Diagnosing cardiac lymphoma in a timely fashion may allow initiation of treatment, enabling a chance for remission or even cure. Primary cardiac lymphoma often presents with additional cardiac symptoms as well as constitutional symptoms and laboratory abnormalities such as elevated lactate dehydrogenase and erythrocyte sedimentation rate [5], all of which may be considered alarming signs. Nevertheless, primary cardiac lymphoma will often be ultimately diagnosed incidentally. Though uncommon, the diagnosis of cardiac tumors in general and lymphoma in particular should be kept in mind in patients presenting with

CEA = carcinoembryonic antigen

arrhythmias, as early diagnosis may be life saving.

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