Left Ventricular Non-Hodgkin Lymphoma Visualized on Contrast Echocardiography

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ABSTRACT: We present a case of AIDS-related Burkitt's type cardiac lymphoma in a middle-aged woman with Epstein-Barr virus infection and profound immunodeficiency. The original features of our case include left ventricular location, female sex, and the use of contrast echocardiography to help establish the diagnosis. © 2007 Wiley Periodicals, Inc. *J Clin Ultrasound* **35**:462–464, 2007; Published online in Wiley Inter-Science (www.interscience.wiley.com). DOI: 10.1002/ jcu.20344

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A⁴¹-year-old woman coinfected with human immunodeficiency virus (HIV), hepatitis B virus, hepatitis C virus, and Epstein-Barr virus was admitted with cauda equina syndrome and a left axillary mass.

One week prior to admission, the patient developed excruciating lower back pain and rectal bleeding and was unable to walk. MRI of thoraco-lumbar spine showed an epidural mass at the level of the fourth lumbar vertebra. She reported that she was not taking highly active antiretroviral therapy therapy for her HIV infection. On admission, her temperature was 37.0°C, her pulse was 100 beats per minute, and her respiratory rate was 20 breaths per minute. Her blood pressure was 159/95 mm Hg. The oral cavity showed presence of thrush. A nontender, hard, palpable, immobile mass in the left axilla measuring 10 \times 10 cm was observed. Muscle weakness was noted on the lower extremities. Patellar deep tendon reflex could not be elicited in either leg. There was decreased sensation over the left inguinal ligament and hyperesthesia on the bottom of the feet.

The patient's last CD4 count was 798 cell/ μ l, and her viral load was 482,558 copies/ml. Contrasted-enhanced CT examination of the chest, abdomen, and pelvis with showed a 10.5 \times 2.8 cm mass in the left axilla, smaller degree of lymphadenopathy in the right axilla, left pleural effusion, splenomegaly, and diffuse abdominal and pelvic lymphadenopathy.

A transthoracic echocardiogram revealed a large highly mobile globular mass in the left ventricle (LV) measuring at least 2.7×1.5 cm in the apical 4-chamber view (Figure 1). It had a broad base of attachment to the endocardium of the anterolateral LV segment at the base of the anterolateral papillary muscle. Using the bolus technique, 0.2 ml of a perflutren lipid microsphere agent (Definity; Bristol-Myers Squibb Medical Imaging, North Billerica, MA) was injected intravenously followed by a 10-ml slow saline flush. On low-mechanical index real-time ultrasound harmonic imaging using a 3.5-MHz harmonic probe connected to a Sequoia scanner (Siemens Acuson, Mountain View, CA), the mass enhanced indicating that it contained its own blood supply. The contrast enhancement was assessed visually; no dedicated software was used. These findings were consistent with a malignant cardiac tumor; given the patient's clinical background, cardiac lymphoma was suspected (Figure 1A).

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INTRACARDIAC LYMPHOMA

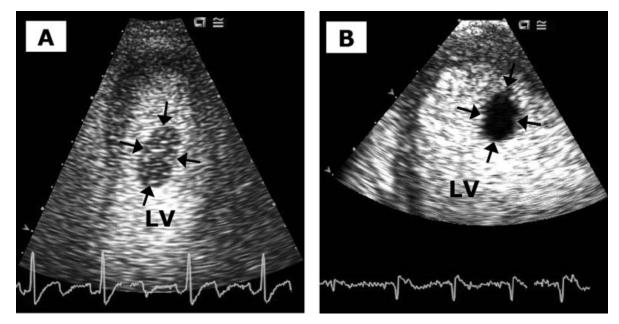


FIGURE 1. The use of intravenous perflutren lipid microbubble echocardiographic contrast helps in the differential diagnosis of an intracardiac mass. (A) Microbubbles are taken up by the cardiac lymphoma (arrows). (B) For comparison, There is no microbubble uptake in a similarly located cardiac thrombus (arrows) in the left ventricle (LV) in a different patient.

The LV wall motion was hyperkinetic, and the LV ejection fraction was increased (\geq 75%). There was mild pulmonary hypertension with a pulmonary artery systolic pressure estimated at 35–40 mm Hg. No pericardial effusion was visualized.

Surgical pathology of the biopsied fourth lumbar vertebral mass and axillary tumor was consistent with malignant lymphoma having highgrade B cell phenotype with a high proliferative index (Ki-67 = 100%) (Figure 2). Neoplastic cells were positive for CD20, CD10, and CD45 and negative for CD3 and BCL-2 markers and carried translocation within the *c-myc* gene. All of these pathologic findings were consistent with Burkitt's lymphoma. Bone marrow biopsy and cerebrospinal fluid analysis were free of malignancy.

The patient received intrathecal chemotherapy and 2 cycles of systemic chemotherapy using the National Cancer Institute EPOCH regimen of etoposide, prednisone, vincristine (Oncovin), cyclophosphamide, and doxorubicin hydrochloride (Adriamycin). A repeat transthoracic echocardiogram only 3 days after initiation of chemotherapy showed a decrease in size of the LV mass to 1.5×1.1 cm. The patient was discharged in good condition and was lost to further follow-up.

DISCUSSION

This case of a woman with an AIDS-related lymphoma embodies the cardinal features of the disorder: prior HIV and Epstein-Barr virus infection, profound immunodeficiency, Burkitt's lymphoma

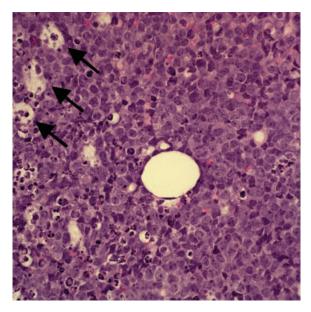


FIGURE 2. Pathologic specimen from the left axillary mass shows typical histologic features of Burkitt's lymphoma. Note the "starry sky" pattern created by the light-colored macrophages (arrows) scattered among the dark lymphoma cells (hematoxylin-eosin stain; original magnification, $40 \times$).

subtype, extranodal involvement including the heart, and poor prognosis.

The original features of our case include the location of the tumor in the left ventricle (most lymphomas occur in the right atrium¹), female sex (there appears to be only 1 prior report of AIDS-related intracardiac lymphoma in a female²), and the use of contrast echocardiogra-

phy to help establish the diagnosis (to our knowledge, this has not been previously reported for a Burkitt's-type intracardiac lymphoma).

Malignancies that are metastatic to the heart are considerably more common than primary cardiac tumors. Lymphoma has the highest relative incidence of cardiac metastases save for melanoma.³ Both metastatic as well as the very rare primary cardiac lymphomas occur more commonly in immunocompromised individuals.⁴

In HIV-infected individuals, the risk of development of systemic non-Hodgkin lymphoma is 60–200 times that of the general population.¹ Cardiac complications in AIDS-related non-Hodgkin lymphoma appear late in the course of illness and in severely immunodeficient state with extremely poor prognosis.⁵

Echocardiography is the primary means of diagnosing cardiac tumors. At present, the major clinical indication for contrast-enhanced echocardiography is endocardial border delineation for the evaluation of global and segmental left ventricular function.⁶ Because malignancies have abnormal neovascularization that supplies rapidly growing tumor cells, often in the form of highly concentrated dilated vessels, such tumors have a high uptake of echocardiographic contrast. On the other hand, fresh thrombi have little or no uptake, because they lack their own blood supply (Figure 1B). The extent of contrast uptake by benign tumors, most of which have a paucity of blood vessels, falls between malignant tumors and thrombi.⁷

Kirkpatrick et al.⁸ have extended the use of this modality to determine the relative perfusion of cardiac masses to help differentiate between a

benign tumor, a malignancy, and a thrombus by using a high-energy ultrasound flash to destroy the contrast microbubbles in the tumor and the surrounding cardiac tissue, then allowing repopulation of contrast bubbles in these structures. This technique decreases the chance of recording a false-positive perfusion and thus mistaking a hypovascular benign tumor or thrombus for a malignancy.

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