Malignant metastases are among the most common cardiac masses. Approximately 9% of metastases to the heart are secondary to malignant lymphomas.1,2 Burkitt lymphoma, although exceedingly rare, can present as an intracardiac mass, most often in the right atrium.3 We report a case of cardiac involvement by Burkitt lymphoma in a 49-year-old man who presented with dyspnea and palpitations. Subsequent diagnostic imaging, pleural fluid cytology, and flow cytometry guided diagnosis and treatment.

In December 2016, a 49-year-old man presented with a 2-month history of dyspnea and palpitations. His relevant medical history included well-controlled human immunodeficiency virus (HIV) infection and Burkitt lymphoma in remission for 3 years after chemotherapy.

At presentation, the patient was experiencing atrial fibrillation with variable ventricular response (40–140 beats/min). Physical examination revealed jugular venous distention, decreased breath sounds in both lung bases, and an extra heart sound at the left lower sternal border consistent with a tumor plop. No lymphadenopathy or abdominal masses were noted. Transthoracic echocardiograms revealed 2 globular masses in the right atrium, one of which partially encased the tricuspid valve, and a thickened myocardium (Fig. 1). A computed tomogram of the chest revealed pleural effusion and a mediastinal lymphadenopathy compressing the right pulmonary artery and superior vena cava. A 1.9 × 1.6-cm filling defect was seen in the posterior right atrium (Fig. 2), which raised suspicions of lymphomatous invasion of the myocardium and pericardium. The images suggested that the dyspnea and conduction abnormalities were due to the progression of Burkitt lymphoma and invasion of the right atrium, myocardium, and pericardium. Results of pleural fluid cytology showed atypical lymphoid cells with enlarged nuclei and prominent nucleoli. Results of flow cytometry confirmed a CD10-positive B-cell population, consistent with Burkitt lymphoma.

The patient was started on an intravenous chemotherapy regimen consisting of rituximab (375 mg/m² on day 1), cyclophosphamide (800 mg/m² on day 1 and 200 mg/m² on days 2–5), vincristine (1.5 mg/m² on days 1 and 8), doxorubicin (40 mg/m² on day 1), and methotrexate (a 300 mg/m² loading dose on day 10 followed by a 2,700 mg/m² infusion over 24 hours). After one cycle of chemotherapy, an echocardiogram showed a marked reduction in the tumor burden (Fig. 3), and the patient’s presenting symptoms had resolved. He continued receiving chemotherapy thereafter, but his cancer status after the first cycle of chemotherapy was unknown to us.
Discussion

Nonmalignant intracardiac masses are typically infections, thrombi, structural abnormalities, and benign neoplasms such as myxomas, lipomas, and angiomas. On the other hand, malignant intracardiac masses are typically breast, lung, or melanoma metastases; primary cardiac tumors are less common. Lymphoma metastases to the heart have an estimated incidence of 9% to 24%. They often occur in immunocompromised patients (for example, HIV-positive patients), but are still unusual. Most lymphoma metastases are diffuse large B-cell lymphomas.

To our knowledge, only 22 cases of intracardiac Burkitt lymphoma have been reported. Most occurred in men. As in our case, the presenting symptoms were usually shortness of breath and palpitations or extra heart sounds; however, these symptoms are nonspecific and difficult to associate with cardiac metastases. The tumor typically involved the right atrium (16 cases, 71%) or the left atrium (5 cases, 23%). Only 2 case reports noted more extensive involvement: in the intra-atrial septum and pulmonary infundibulum in one case, and in the right and left atria, right ventricle, and tricuspid valve in the other. Our patient’s case is unique because the metastatic lymphoma caused substantial myocardial infiltration and pericardial disease. Such extensive involvement by a rare intracardiac tumor has not been reported.

Our case highlights the importance of cardiac imaging in diagnosing systemic diseases and in monitoring the response to treatment. Echocardiography was performed early because of the patient’s arrhythmia and associated dyspnea. The echocardiographic evidence
of extensive cardiac tumor and the patient’s history of Burkitt lymphoma prompted us to obtain a CT scan of the chest and analyze pleural fluid. A follow-up echocardiogram obtained after one cycle of chemotherapy showed a substantial reduction in intracardiac tumor burden.

References

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