primary Sjogren syndrome (pSS) is an autoimmune condition characterized by exocrine gland involvement resulting in dryness of the eyes and mouth and extraglandular manifestations. It is associated with a 44-fold increased risk for the development of non-Hodgkin lymphoma. Lymphoma develops in approximately 4% of patients with pSS, which manifests in diverse nodal and extranodal sites such as the salivary glands, stomach, lung, and adrenal glands. This would be the first reported case of pSS-related cardiac lymphoma. The lymphoma was initially confined to the heart but rapidly progressed. The importance of early appropriate radiologic and surgical intervention is highlighted.

Clinical Summary
An 84-year-old Chinese woman presented with increasing dyspnea associated with weight loss. She had mild arthritis and dryness of the eyes and mouth for approximately 10 years. She was a nonsmoker and had no family history of cardiac illnesses. On examination, the heart sounds were muffled and jugular venous pressure was increased. Systemic examination results were otherwise unremarkable. The results of investigations were as follows (reference ranges in parentheses): Hemoglobin was 9.6 g/dL (12.0–16.0); white cell count and platelet count were normal. Erythrocyte sedimentation rate was 96 mm/h (3–20), and C-reactive protein was 26.3 mg/L (0.2–8.8). Schirmer’s I test was positive at 2 mm (<5 mm). Anti-Ro and La antibodies were present. Anti-dsDNA, Smith, ribonucleoprotein, ScI-70, and cardiolipin antibodies were not present. Serum complements C3 and C4, urea and electrolytes, liver enzymes, bone and thyroid profile, rheumatoid factor, and immunoglobulins G, A, and M were unremarkable. A computed tomographic (CT) scan and an echocardiographic scan showed a large pericardial effusion, with no other abnormality detected. Needle pericardiocentesis yielded nonhemorrhagic pericardial fluid that was negative for malignant cells and organism growth. Needle pericardiocentesis yielded nonhemorrhagic pericardial fluid that was negative for malignant cells and organism growth. Needle pericardiocentesis yielded nonhemorrhagic pericardial fluid that was negative for malignant cells and organism growth. Needle pericardiocentesis yielded nonhemorrhagic pericardial fluid that was negative for malignant cells and organism growth. Needle pericardiocentesis yielded nonhemorrhagic pericardial fluid that was negative for malignant cells and organism growth. Needle pericardiocentesis yielded nonhemorrhagic pericardial fluid that was negative for malignant cells and organism growth.

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Cardiac lymphoma in primary Sjogren syndrome: A novel case established by targeted imaging and pericardial window

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and bcl2, and demonstrated a proliferation index of 80% with Ki67 but were negative for CD3 and cytokeratin (MNF116). The features were consistent with CD20-positive diffuse large B-cell lymphoma (DLBCL). At this point, our patient declined chemotherapy because of frailty. Subsequent clinic review showed new findings of enlarged parotid glands and a left breast mass. A CT scan performed 4 weeks after the diagnosis of primary cardiac lymphoma revealed progression of disease with the involvement of intra-abdominal nodes and left breast but no lung involvement. Bone marrow examination was unremarkable. Biopsy of the breast mass confirmed CD20-positive DLBCL.

Discussion

DLBCLs are aggressive tumors, and the early commencement of a chemotherapy regimen that includes rituximab yields better outcomes. With the patient’s eventual consent, treatment comprising rituximab, cyclophosphamide, vincristine, and high-dose prednisone was initiated. After 4 courses of chemotherapy, there has been clinical improvement with return of normal appetite, weight gain, and no breathlessness or chest pain. Follow-up CT scans of the neck, thorax, and abdomen suggest good response to treatment with reduction of abnormal thickened pericardium and almost complete resolution of widespread lymphadenopathy.

Cardiac lymphoma is rare and tends to occur in human immunodeficiency virus-positive individuals. Our patient had pSS but was human immunodeficiency virus negative. Primary cardiac lymphoma may be low grade; however, as highlighted in this brief communication, it may be insidious but aggressive and occur in preexisting nononcologic syndromes, such as pSS. Diagnosis of cardiac lymphoma requires a high index of suspicion. Echocardiographic evidence of clinically silent cardiac involvement in pSS is common. However, Mita and colleagues found that 21.2% of patients with pSS had pericardial effusions that rarely caused symptoms.

In patients with pSS with symptomatic pericardial effusions, prompt evaluation with cardiac magnetic resonance imaging or positron emission tomographic scans for lymphoma should be considered because of significant disease association that may not be initially evident on echocardiography or pericardial fluid analyses. Urgent pericardial window creation and biopsy of the potentially occult plaque-like cardiac lesion for therapeutic and diagnostic purposes in our patient were imperative for optimal outcome.

References