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A Rare Case of Lymphocytic Lymphoma with Pericardial Infiltration

B. Mesmoudi[1], J. Nguadi[1], R. Jennane[1], M.Ababou[2], N.Loudiyi[1], I.Asfalou[1] and A.Benyass[1]

[1] Department of Cardiology, Military Hospital Mohammed V of Rabat, University Mohammed V of Rabat

[2] Department of Hematology, Military Hospital Mohammed V of Rabat, University Mohammed V of Rabat

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ABSTRACT

Small lymphocytic lymphoma (SLL) is a low-grade B-cell non-Hodgkin lymphoma (NHL) characterized by a monoclonal population of mature B-cell lymphocytes. It is rarely associated with cardiac involvement. Pericardial effusion is one of the most common incidental findings in cancer patients, which significantly worsens morbidity and mortality. Thus, early detection, adequate treatment, and understanding of pericardial diseases are necessary to help improve the quality of life of cancer patients. An individual treatment plan should be established, taking into account cancer stage, the patient's prognosis, local availability and experience. We report the case of a SLL involving the pericardium.

Introduction

Secondary or metastatic cancers of the pericardium account for 75% of all cardiac malignancies[1]. Small lymphocytic lymphoma (SLL) is a low-grade B-cell non-Hodgkin lymphoma (NHL) that usually presents with lymphadenopathy. It is rarely associated with cardiac involvement[2]. The pericardium, including the epicardium, is the most common location of cardiac involvement by secondary tumors. Pericardial effusion, either due to cancer invasion or cancer treatment, is one of the most common incidental findings in cancer patients, which significantly worsens morbidity and mortality. Thus, early detection, adequate treatment, and understanding of pericardial diseases are necessary to help improve the quality of life of cancer patients. We report the case of a SLL involving the pericardium.

Case report

We report the case of a patient aged 60 years, diabetic under medical treatment, treated for lymphocytic lymphoma since June whose diagnosis was made by scanno-guided biopsy of a left para-aortic adenopathy with immunohisto chemistry : CD20+, CD5+, CD23+, cyclin D1 -, CD10-, Bcl2+. The G-CIRS score was at 07.

The extension evaluation showed on the chest X-ray: a filling of the left pleural cul-de-sac. He underwent a PET scan on June (*Figure 1A*) which revealed: Numerous pathological hypermetabolic lymph node above and below the diaphragm at the following levels: bilateral cervical, right axillary, mediastinal, abdominal and probably left intercostal, with SUVmax ranging from 2.3 to 13.5. No other suspected pathological hypermetabolism in the rest of the body explored from head to mid-thigh was found. There was a bilateral pleural effusion, mostly on the left, non-fixing. Transthoracic echocardiography performed as part of the pretherapy assessment was normal, and the ventricular ejection fraction was 65%. Therapeutically, the patient was put on Corticotherapy 1mg/kg and the decision of the staff was to treat him with FCR cure (Fludarabine, Cyclophosphamide, and rituximab). We received the patient four months later in the emergency room for a stage IV of NYHA dyspnea. The patient underwent a chest X-ray showing a bilateral pleural effusion of moderate size. On physical examination the patient was hemodynamically stable with a blood pressure at 110/64 mmHg, with a heart rate of 90 bpm. On auscultation, heart sounds were normal with no pericardial friction rub. On echocardiography, a left ventricular hypertrophy of infiltrative appearance was noted, associated with a pericardial infiltration, with nodularity, and a pericardial effusion of moderate size (Figure 2). A complementary cardiac MRI was done confirming the diagnosis and eliminating an intra-cardiac mass. A control PET scan was done, showing a cardiac fixation previously absent on the PET scan dating from 4 months ago (*Figure 1B*). The patient was hospitalized in clinical hematology for further management.

Discussion

SLL is the equivalent of chronic lymphocytic leukemia (CLL) that occurs in the peripheral blood and is characterized by a monoclonal population of mature B-cell lymphocytes 2. Because of its indolent course, SLL is often diagnosed incidentally. The symptoms of SLL typically range from constitutional symptoms, such as night sweats, weight loss, and fatigue, to symptomatic enlargement of lymphoid tissue 3. Cardiac involvement in SLL is extremely rare, with pericardial involvement being exceedingly rare. In a retrospective analysis of 94 patients with cardiac involvement of non-Hodgkin's lymphoma from 1990 to 2015, Gordon et al.4 ;5 found that SLL was the underlying malignancy in six cases (7%). The presenting symptoms for these cases were heart failure (four cases), arrhythmia (one case), and cardiac arrest (one case).

Malignant pericardial effusion is a common and serious manifestation in malignancies. Despite the monumental advances in the diagnoses and therapeutics of malignancy, several cancer patients have presented with pericardial involvement, including acute pericarditis, constrictive

Tele:

E-mail address: bouthayna.mes@gmail.com

pericarditis, and pericardial effusion. Pericardial metastases occur late in the course of a neoplasm, usually as recurrent disease. The median time of onset is 20 months after initial diagnosis. Rarely, a malignant pericardial effusion is the first sign of a neoplasm, and that is why malignancy must be excluded in every case of an acute pericardial disease with cardiac tamponade at presentation, rapidly increasing pericardial effusion and an incessant or recurrent course. Thus, the definite differentiation of malignant pericardial effusion and rapid diagnosis have therapeutic and prognostic importance3. In a patient with a malignant tumor and pericardial effusion, the differential diagnosis includes not only malignant pericardial disease but also benign idiopathic, drug-induced, or radiation induced pericarditis. Pericardial effusion can be due to several conditions, such as cell infiltrates from extramedullary hematopoiesis, hemorrhagic diathesis due to thrombocytopenia, and in patients receiving chemotherapy pericardial effusion may be a side effect of treatment, or could be a result of an infection 6;7. Malignant pericardial effusion can lead to various adverse events, including chest discomfort, dyspnea, tachycardia, hypotension and cardiogenic shock, although some patients remain undiagnosed until death. The patient may have a mild presentation, as is often seen in the early stage of pericardial effusion, or may present with dramatic hemodynamic compromise, as in cardiac tamponade, which is a life-threatening emergency. Impairment of cardiac function occurs in approximately 30% of patients and is usually attributable to pericardial effusion 8. Metastases in lymphomas are usually focal, firm nodules that may be found in the walls of all cardiac chambers and in the pericardium9.Once suspected, we should identify the cause of the pericardial effusion. A low QRS voltage and electrical alternans on EKG might be present. Metastatic involvement of the pericardium has non specific imaging findings and is often suggested by the diagnosis of the primary non cardiac neoplasm. Imaging findings include presence of effusion, irregular thickening of the pericardium or nodularity, and distinct pericardial masses10. Echocardiography is the primary imaging modality for pericardial effusion and has a central role in the disease evaluation, follow-up and therapy. The size of the effusion needs to be monitored through regular echocardiography. Echocardiography should be repeated within one month in those with small effusions, two weeks in moderate effusions, and within a week in those with large effusions 11. Echocardiographic evaluation is essential in oncological patients before, during and after the end of treatment3. Computed tomography (CT) and cardiac magnetic resonance (CMR) are valuable imaging tools in the evaluation of pericardial disorders 12,13. CT can provide initial characterization of pericardial fluid in patients with pericardial effusion of unknown etiology. It allows also the search for the possible presence of a concomitant thoracic neoplasm. CMR has a superior ability to characterize pericardial effusions and masses14. Both CT and MRI are indicated for suspected localized or hemorrhagic effusion, pericardial thickening, or effusive-constrictive pericarditis.

Most patients presenting with malignant pericardial effusion have a poor prognosis. The available treatment options vary from simple drainage to surgery, with the primary aims of relieving the symptoms and improving quality of life. Doses of cancer drugs may be reduced or withheld in chemotherapy-related pericardial effusion based on the prognosis of cancer11.

Management of these patients is multidisciplinary, requiring a team work between cardiologists, oncologists, radiotherapists, palliative care physicians and surgeons. An individual treatment plan should be established, taking into account cancer stage, the patient's prognosis, local availability and experience.

Conclusion

Lymphomatous pericardial effusion is rarely associated with lymphocytic lymphoma (SLL), which is a low-grade B-cell non-Hodgkin lymphoma (NHL). Management of these patients is multidisciplinary, requiring a team work, but there is still need for further research.

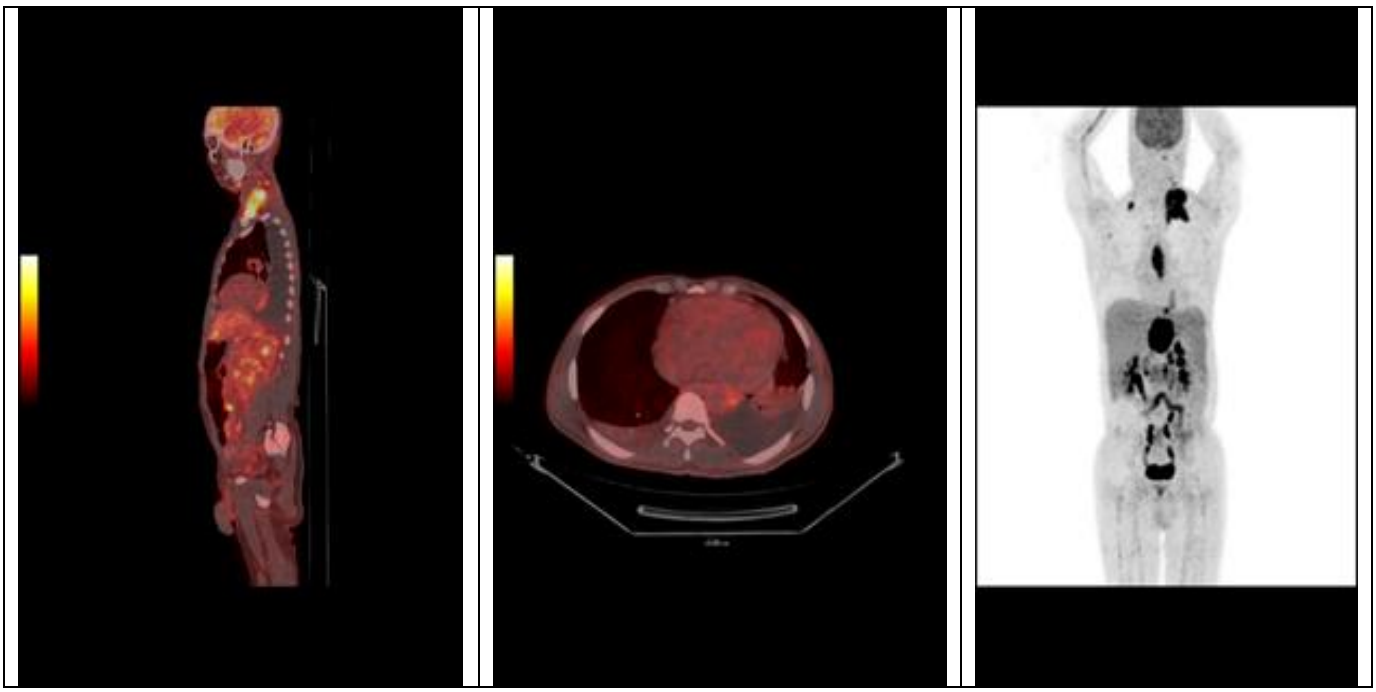

Figure 1 A. PET SCAN before treatment

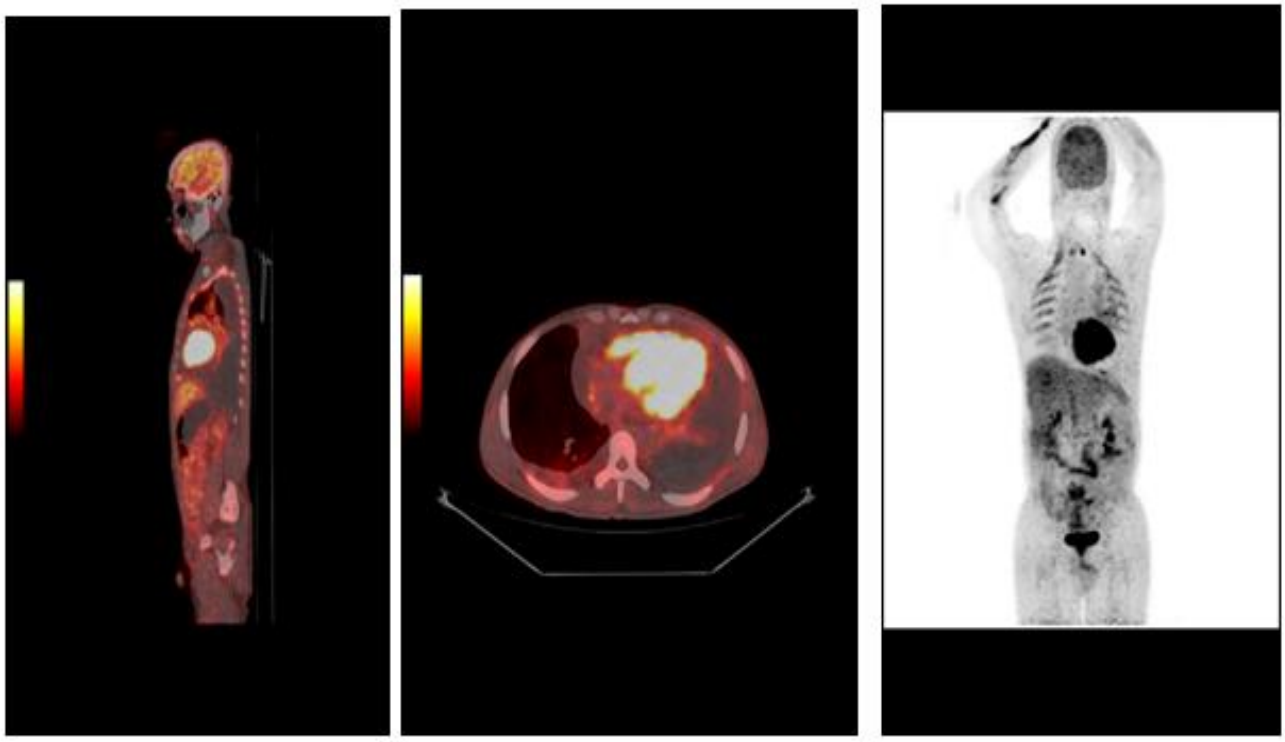

Figure 1B . PET SCAN after treatment

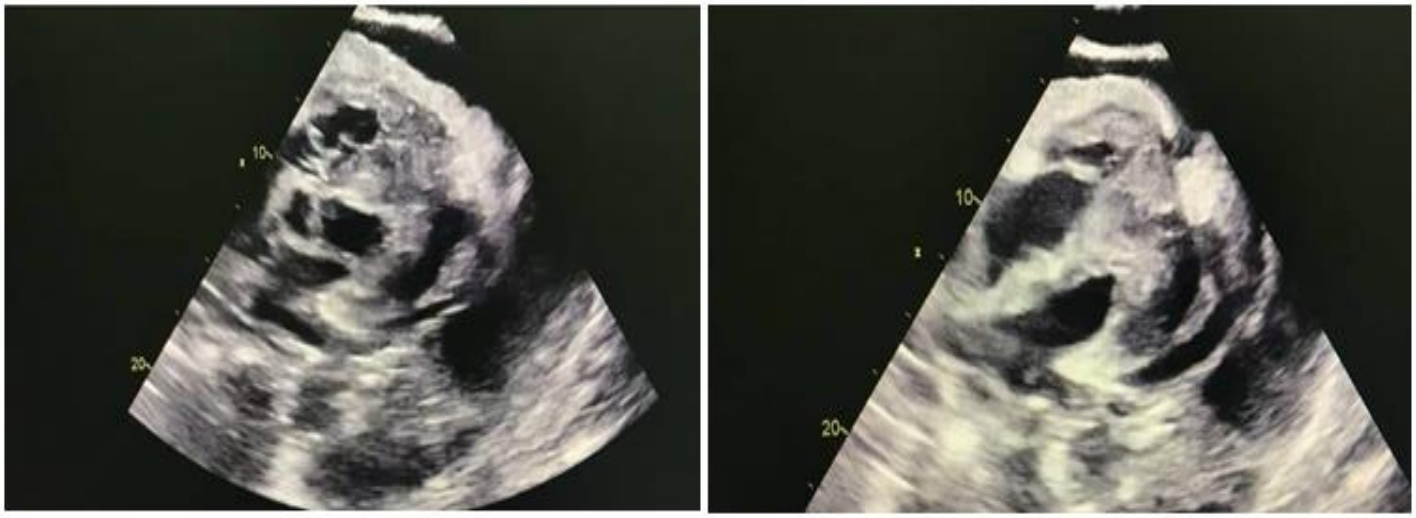

Figure 2. Echocardiography images showing a left ventricular hypertrophy of infiltrative appearance associated with a pericardial infiltration, with nodularity, and a pericardial effusion of moderate size

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