

# A Case of Low-Grade Primary Cardiac Lymphoma with Pericardial Effusion Diagnosed by Combined <sup>18</sup>F-Fluorodeoxyglucose Positron Emission Tomography and Computed Tomography (FDG-PET/CT) Imaging and Effusion Cytology

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Study Design A  
Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
Literature Search F  
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**Conflict of interest:** None declared

**Patient:** Female, 72  
**Final Diagnosis:** Primary cardiac lymphoma  
**Symptoms:** Cardiac tamponade • dyspnea  
**Medication:** —  
**Clinical Procedure:** FDG-PET/CT scan  
**Specialty:** Nuclear Medicine

**Objective:** Rare disease  
**Background:** Primary cardiac lymphoma is rare and can be an aggressive disease, depending on the grade. A case is reported of low-grade primary cardiac lymphoma associated with a pericardial effusion. <sup>18</sup>F-fluorodeoxyglucose positron emission tomography and computed tomography (FDG-PET/CT) imaging was useful in the diagnosis and in evaluating the disease activity in this case.

**Case Report:** A 72-year-old Japanese woman visited a general practitioner, complaining of dyspnea associated with cardiac tamponade. Pericardiocentesis was performed, and Group V malignant cells were identified by cytology, suspicious for malignant lymphoma. Whole-body FDG-PET/CT scans showed no pleural effusion or lymph node metastasis supporting the diagnosis of primary cardiac lymphoma diagnosed on pericardial effusion. The laboratory investigations showed that levels of serum soluble interleukin-2 (IL-2) receptor (sIL-2R), a diagnostic and prognostic marker for malignant lymphoma, were not elevated (258 U/ml). A six-month follow-up FDG-PET/CT scan showed an increased volume of the pericardial effusion and mild but abnormal uptake diffusely in the pericardial space, and the sIL-2R was slightly elevated (860 U/ml). No abnormal FDG accumulation outside the retained pericardial effusion was noted, which was compatible with a clinical picture of low-grade primary cardiac lymphoma, and in a period of watchful waiting during the first two years later, the sIL-2R had reduced to 195 U/ml.

**Conclusions:** This is a rare case of low-grade primary cardiac lymphoma detected in a pericardial effusion, and highlights the utility of the FDG-PET/CT scan as a valuable diagnostic and follow-up modality.

**MeSH Keywords:** Adenolymphoma • Fluorodeoxyglucose F18 • Heart Neoplasms • Positron-Emission Tomography

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## Background

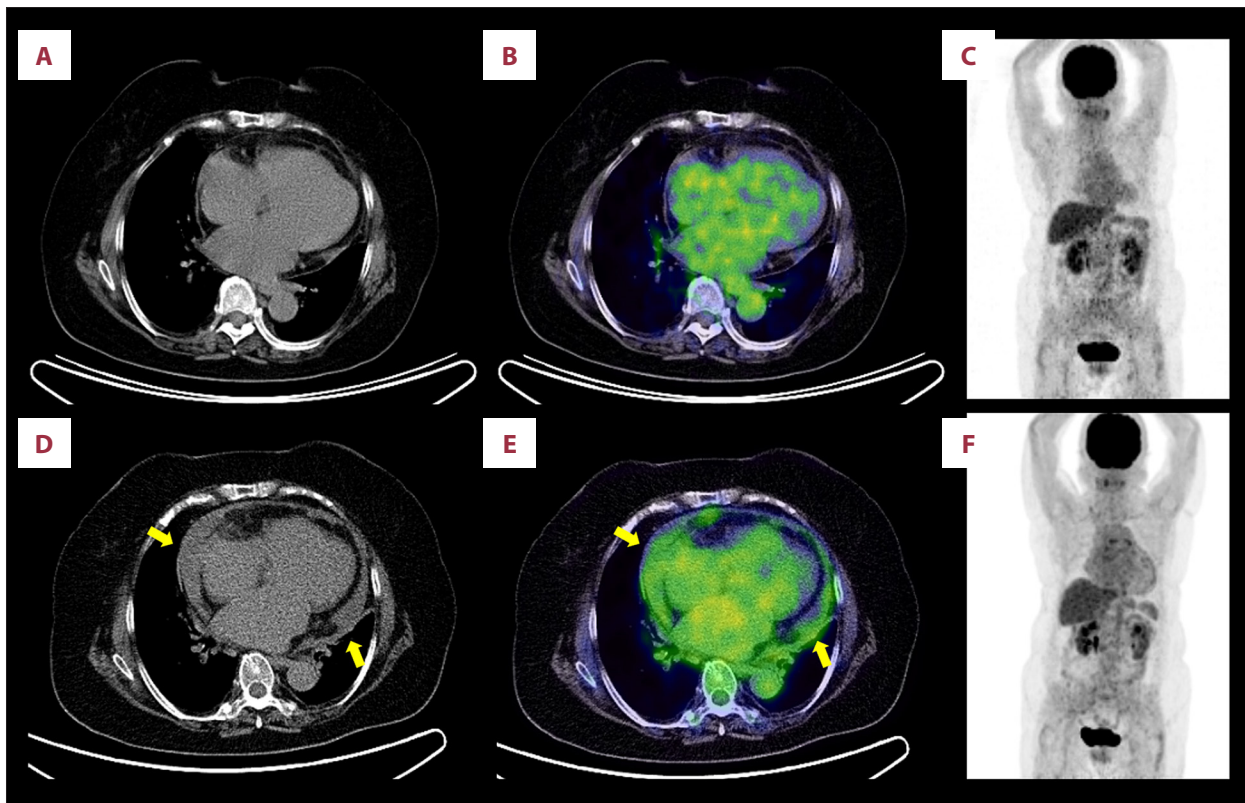
Primary cardiac lymphoma is an exceptionally rare tumor and, depending on the grade of lymphoma, can be fatal with an aggressive course [1]. Primary cardiac lymphoma is more likely to arise in the right side of the heart and epicardium and is most commonly reported to be a diffuse large B-cell non-Hodgkin's lymphoma. Although pericardial effusion or pericardial thickening is often a common early feature of primary cardiac lymphoma, it has non-specific imaging features that result in a diagnostic challenge in the absence of a biopsy diagnosis, such as a bulky or infiltrative mass, for example, atrial or ventricular wall infiltration with extension along the epicardial surface [2]. Pericardial effusion cytology may be the most common procedure for the diagnosis of primary cardiac lymphoma, but invasive pericardiocentesis is not routinely performed unless the patient has symptoms due to the pericardial effusion. Diagnostic evaluation and disease activity is important to the appropriate grading

and staging of primary cardiac lymphoma, as well as its early treatment [3].

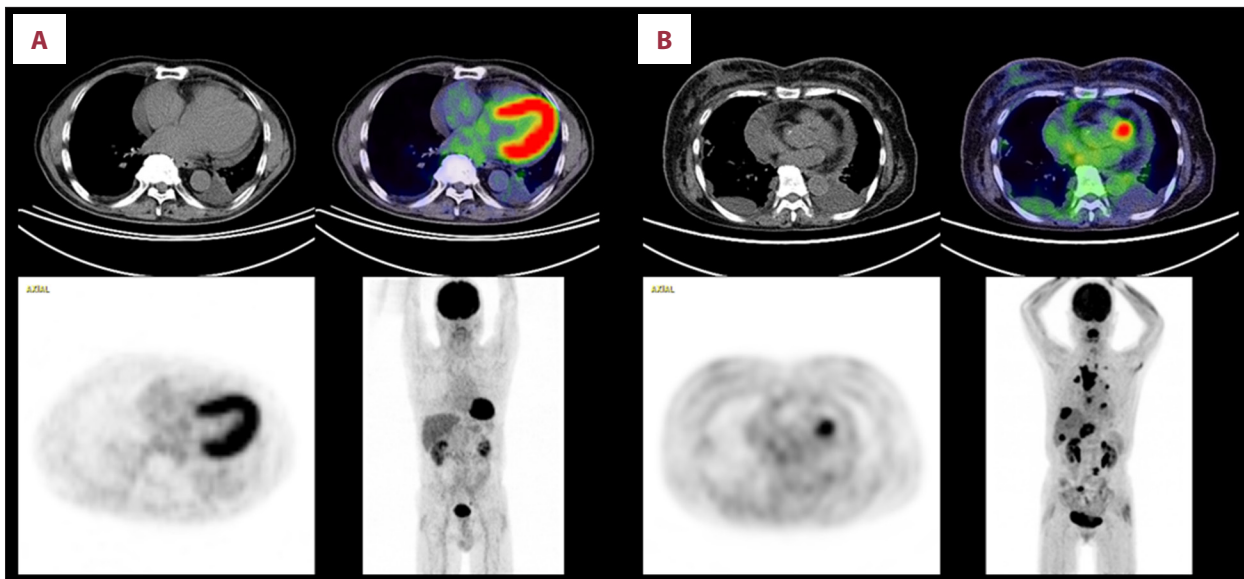
This report is of a case of low-grade primary cardiac lymphoma with pericardial effusion diagnosed by pleural fluid cytology, in which the specific findings of abnormal pericardial uptake using  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography and computed tomography (FDG-PET/CT) imaging was a useful approach when making the diagnosis and in evaluating the disease activity on follow-up.

## Case Report

A 72-year-old Japanese woman visited a general practitioner, with the chief complaint of dyspnea. A clinical diagnosis of pericardial effusion was made, and she was referred to a district hospital. Pericardial effusion was confirmed and cardiac tamponade was shown on enhanced computed tomography (CT)



**Figure 1.**  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography and computed tomography (FDG-PET/CT) imaging. (A, D) Cardiac computed tomography (CT) imaging. (B, E)  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography and computed tomography (FDG-PET/CT) fusion imaging. (C, F) Maximum intensity projection of a whole-body of FDG-PET imaging. The first FDG-PET examination (upper row) shows no pericardial effusion and no lymph node metastases. The second FDG-PET/CT examination was undergone six months after the first visit (lower row). The  $^{18}\text{F}$ FDG uptake of the pericardial space using region of interest (ROI) diameter of 20 mm was measured and the maximum standardized uptake value ( $\text{SUV}_{\text{max}}$ : maximum ROI activity/[injected dose/body weight], with activity in megabecquerel [MBq] per gram, dose in MBq and weight in grams) was calculated. FDG-PET/CT showed an increased volume of the pericardial effusion and diffuse mild  $^{18}\text{F}$ -FDG uptake ( $\text{SUV}_{\text{max}} 2.7$ ) corresponding to the retained pericardial effusion (D, E) and no lymph node metastasis (F).



**Figure 2.** Representative cardiac  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography and computed tomography (FDG-PET/CT) images. **(A)** Pericardial and pleural effusions in congestive heart failure. Note that no or less uptake of  $^{18}\text{F}$ -FDG in the pleural effusion associated with heart failure when compared with primary cardiac lymphoma (Figure 1E). **(B)** An example of carcinomatous pericarditis and cardiac metastases from a case of lung cancer. Focal and high  $^{18}\text{F}$ -FDG uptake was observed in the left myocardium, and multiple areas of mild uptake in the pericardium were shown corresponding to cardiac metastases with carcinomatous pericarditis. A primary right lung cancer and multiple distant metastases can be identified as hot spots in the FDG-PET whole-body scan.

imaging. Following a physical examination, a pericardiocentesis was performed, and approximately 1,000 ml of bloody fluid was drained from the pericardial space. Group V malignant cells were identified by cytology, suspicious of malignant lymphoma including diffuse B-cell non-Hodgkin's lymphoma [4]. The patient was then referred to our university hospital for further examination and treatment.

Whole-body FDG-PET/CT imaging scans were undertaken with a Biograph Duo or Biograph 40 PET/CT system (Siemens AG, Erlangen, Germany) using the following standard clinical protocols. The patient fasted for a minimum of four hours, and 185.0–370.0 megabecquerel (MBq) (5.0–10.0 mCi) of  $^{18}\text{F}$ -FDG was administered by intravenous injection. The patient was rested for one hour between tracer injection and commencing PET/CT image acquisition. A blood glucose concentration at the time of the FDG-PET/CT scan was below 100 mg/dL. FDG-PET/CT showed no pleural effusion or lymph node metastasis, supporting the diagnosis of primary cardiac lymphoma (Figure 1A–1C). The laboratory investigations showed that levels of serum soluble interleukin-2 (IL-2) receptor (sIL-2R), a diagnostic and prognostic marker for malignant lymphoma, were not elevated (258 U/ml).

Six months after the patient's first hospital visit, she was free from symptoms, including dyspnea, and the residual fluid from pericardial effusion was not large enough to require further

pericardiocentesis. However, the FDG-PET/CT scan were sensitive enough to show mild but abnormal uptake in the pericardial space (Figure 1D–1F), and the sIL-2R was slightly elevated (860 U/ml). The  $^{18}\text{F}$ -fluorodeoxyglucose ( $^{18}\text{F}$ -FDG) uptake of the pericardial space, using region of interest (ROI), was measured and the maximum standardized uptake value ( $\text{SUV}_{\text{max}}$ ) was 2.7. This case showed no remarkable  $^{18}\text{F}$ -FDG accumulation outside the retained pericardial effusion, being compatible with a clinical picture of low-grade primary cardiac lymphoma [5]. In a period of watchful waiting during the first two years after diagnosis, the sIL-2R level was 195 U/ml.

## Discussion

Primary cardiac lymphoma can be associated with varied symptoms and signs, including respiratory distress, pericardial effusion, and cardiac tamponade, and can also result in cardiac failure [1].

Although retained pericardial effusion is a particularly common finding in cardiac failure, it does not consist of an accumulation of  $^{18}\text{F}$ -FDG (Figure 2A). Other potential differential diagnoses include myocardial metastasis, carcinomatous pericarditis, and cardiac inflammatory diseases (Figure 2B). No abnormal  $^{18}\text{F}$ -FDG accumulation was observed outside the retained pericardial effusion in this case. It is unlikely that a metastatic tumor and, in

particular, carcinomatous pericarditis could be demonstrated as solitary abnormal uptake on  $^{18}\text{F}$ -FDG/PET, without the presence of the most frequent primary lesions, including lung cancer or breast cancer, and other distant metastases. Furthermore, a diffuse distribution pattern of  $^{18}\text{F}$ -FDG to the retained pericardial effusion, not localized to the pericardium or endocardium, could be a characteristic finding of primary cardiac lymphoma corresponding to viable lymphoma cells floating in the pericardial fluid. Inflammatory pericarditis with mild diffuse  $^{18}\text{F}$ -FDG uptake in the sites of pericardium or pericardial effusion can be difficult to distinguish from primary cardiac lymphoma, but the clinical presentation including fever, chest pain, and the presence of a cardiac murmur could be a clue to diagnosis.

Previous case reports of primary cardiac lymphoma with fatal outcomes have been reported, but cases of remission following chemotherapy have also been reported [6,7]. Currently, established diagnostic imaging approaches, with FDG-PET/CT including the use of the maximum standardized uptake value ( $\text{SUV}_{\text{max}}$ ) and cut-off values for the differentiation of cardiac

tumors are limited. However, a diffuse  $^{18}\text{F}$ -FDG uptake solitary in pericardial effusion has been shown to be key for the diagnosis of primary cardiac lymphoma, in particular, for diffuse large B-cell lymphoma [3]. As this case has shown, early diagnosis and follow-up of primary cardiac lymphoma by the FDG-PET/CT imaging findings are likely to be clinically significant.

## Conclusions

Primary cardiac lymphoma is very rare and may be particularly difficult to diagnose if it is only associated with a pericardial effusion. To the authors' knowledge, this is the first report to demonstrate the early diagnosis and clinical follow-up of a patient with low-grade primary cardiac lymphoma using  $^{18}\text{F}$ -FDGPET/CT imaging.

## Conflicts of interest

None.

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