Cardiac lymphoma: prevalence in non Hodgkin disease and PET-CT FDG uptake pattern.

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Learning objectives

- To review the prevalence of heart involvement in lymphomas encountered in 2-[-fluorine18] fluoro-2-deoxy-D-glucose (FDG) PET/CT as well as the different patterns of FDG cardiac uptake in those cases.
- To correlate findings in PET/CT with other imaging techniques such as contrast enhanced computed tomography (CECT), magnetic resonance (MR), transthoracic echocardiography (TTE) and also with the disease follow up.
- To be aware of the physiological patterns of cardiac FDG activity that can appear in oncologic PET/CT studies.

Background

Cardiac lymphoma (CL) as secondary extension of Non Hodgkin´s Lymphoma (NHL) is described in 30% of autopsies [9], however few cases are diagnosed in vivo.

Primary lymphoma of the heart and pericardium is rare, accounting for 1.3% of primary cardiac tumors and only 0.5% of extranodal lymphomas at autopsy [2-3].

Cardiac symptoms related to lymphoma (dyspnea, congestive heart failure, pericardial effusion and atrioventricular block) are nonspecific and consequently CL is difficult to suspect [1], whereas it is an oncologic emergency due to its rapid evolution and advanced stage of organ infiltration [4-5].

Images of cardiac lymphoma are unspecific in CECT; the most common finding is an ill-defined infiltrative mass in the atrial septum with or without spreading into the atria and less often into the left ventricle [1]. Pericardium might also be affected due to contiguous malignancy of the mediastinum. Image features as focal and firm nodules in the cardiac walls, are considered to be a late manifestation of the disease [10]. Attenuation of these masses in CECT may mimic normal myocardium, so many cases might go undiagnosed.

Integration of PET/CT into routine oncologic imaging has widely improved the assessment of lymphoma in baseline staging and facilitated functional evaluation of disease behavior, metabolic response to therapy, and earlier detection of disease recurrence [6-7].

It is important to understand the range of normal myocardial FDG PET/CT activity in order to be able to recognize the pathologic patterns. We have to be aware of the variable and non-uniform cardiac FDG uptake in oncologic patients as this is why the heart and
paracardiac areas are usually miss-evaluated during interpretation of PET/CT images. Nevertheless the main physiological patterns are broadly described and can be defined as either absent (residual cardiac blood pool), diffusely increased, focally increased (papillary muscles), or regionally increased [8] (figure 1).

Fig. 1: Normal patterns of myocardial FDG uptake. Non-fused PET coronal images. (a) Absent. (b) Basal ring. (c) Focally increased uptake in papillary muscle. (d) Diffusely increased biventricular uptake.

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Findings and procedure details

We reviewed 141 PET/CT initial staging of lymphoma studies that had been performed at our hospital between January 2012 and November 2013. In one case the baseline study was a whole body CECT, so the patient underwent his first FDG PET/CT after 3 courses of chemotherapy. Among the total, 92 were found to be NHL (65%) and 49 Hodgkin’s Lymphoma (35%), 60 women and 81 men with a mean age of 58 (from 15 years old to 85 years old). Fifty cases (35%) had a stage III or higher of the Ann Arbor Staging Modified System for Hodgkin’s and Non- Hodgkin’s Lymphoma at the initial staging.

FDG cardiac activity was non-pathological in 136 patients, the majority showing residual cardiac blood pool activity, followed by a basal ring pattern as a physiological FDG uptake. Other normal patterns were also observed.

Seven cases (5%) revealed a pathological FDG metabolic activity of the heart, with atrial septum involvement the most frequent finding (43%), followed by the infiltration of the pericardium due to contiguous mediastinal bulk (29%). In 14% the left atrium was affected and in one case (14%) lymphoma infiltrated the left ventricle with multiple masses in the ventricular septum in several segments.

The findings were correlated with other imaging techniques such as CT, MR, TTE, and were consistent with the follow-up studies. Pathologic lab works disclosed in all seven cases large B cells (LBC) NHL.

Case 1 (figures 3, 4 and 5)

- Epidemiologic data: 61 year old male.
- Symptoms: weight loss of 10kg in the last 2 months. Non cardiac symptoms registered.
- Diagnosis: LBC NHL, stage IV B, IPI (International Prognostic Index) 3 (intermediate- high risk).
- Treatment: 6 courses of cyclophosphamide, vincristine, liposomal doxorubicin, prednisone and rituximab (R- COMP)

1. Initial staging PET/CT shows an abnormal increase of FDG metabolism in multiple lymph nodes located in cervical spaces, anterior mediastinum and mesentry and extranodal extension to the interatrial septum, gastric fundus, left suprarrenal gland, both kidneys and small bowel (figure 3).
2. There is no pathological FDG uptake after full treatment (figure 5, b).
3. MR after 3 courses of chemotherapy proved a lightly decreased interatrial hyperintense mass. After injection of gadolinium there is a visible interatrial
mass enhancement and a thin linear enhancement within the walls of both left and right atria (figure 4, b, c).

**Fig. 3:** 61 year old male with LBC NHL stage IVB. Fused PET/CT images. (a and b) Axial images show an hypermetabolic mass in the interatrial septum extending to the right atrium. (c) Multiple bilateral renal masses. (d) Small bowel infiltration. (e) Coronal image show interatrial and renal dissemination.

**References:** HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

**Fig. 4:** (a) Fused PET/CT axial image show pathologic interatrial hypermetabolic focus. (b) Axial double IR fat saturation T1 weighted MR depicts an hyperintense mass
in the interatrial septum. (c) Gadolinium enhanced axial double IR fat saturation T1 weighted MR shows malignant enhancement of the mass and the atrial wall.

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

Fig. 5: Fused PET/CT axial images. (a) Interatrial lymphoma. (b) Same level as (a) after treatment shows residual cardiac blood pool activity with non pathologic uptake.

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

Case 2 (figures 6 and 7)

- Epidemiologic data: 79 year old female.
- Symptoms: right breast pain, enlargement and swelling with right arm pain.
- Diagnosis: LBC NHL, stage IV, IPI 2 (low-intermediate risk).
- Treatment: 6 courses of cyclophosphamide, vincristine, prednisone and rituximab (R- COP)

1. CECT depicts a large interatrial mass extending intracavitarily to the right atria and ventricle. We can see bilateral pleural effusion and subcutaneous infiltration of the right breast (figure 6).
2. PET/CT performed after 3 courses of R- COP, shows a remarkable decrease of the interatrial mass (figure 7, b). Note that due to its FDG hypermetabolism and the evident response to chemotherapy, PET/CT enables us to differentiate it from other frequent intracavitary masses with no metabolic activity such as thrombus.
**Fig. 6:** 79 year old female. LBC NHL stage IV. CECT. (a and b) Axial image of the large interatrial mass extending inside the right atria and ventricle. Bilateral pleural effusion and subcutaneous infiltration of the right breast in (a). Coronal (b) and sagittal (c) reconstructions show intracavitary mass inside the right atrium.

**References:** Department of Radiology, Hospital Manacor- Palma de Mallorca /Spain
Fig. 7: (a) CECT axial image shows the interatrial mass occupying right cardiac cavities. (b) Fused PET/CT axial image at the same level as (a) proves hypermetabolic interatrial focus related to residual lymphoma.

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

Case 3 (figure 8)

- Epidemiologic data: 40 year old male. HIV + (recent).
- Symptoms: weakness, weight loss of 20kg, low fever and sweat for one year.
- Diagnosis: LBC NHL, stage IV B, IPI 4 (high risk).
- Treatment: 1 course of cyclophosphamide, doxorubicin, vincristine and rituximab (R-CHOP) + 2 courses of dose-adjusted etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin (DA- EPOCH) + 2 courses of hyperfractionated rituximab- cyclophosphamide, vincristine, doxorubicine, dexametasone / cytarabine, metotrexate (R-HyperCyVAD/R-AM).

1. Initial staging PET/CT describes notable FDG hyperactivity in the enlarged right pectoral muscles, hepatic focal masses, striking homogeneous testicular uptake, multiple spinal focus and a cardiac hypermetabolic focal lesion located in the interatrial septum (figure 8, a, b and c).
2. Intermediate PET/CT after treatment (figure 8,d) demonstrates regression of the interatrial, and bone extension with persistence of the muscular bulk, which is mildly smaller.
Fig. 8: 40 year old male, HIV +, LBC NHL stage IV. (a) FDG hyperactivity within the thoracic wall and interatrial septum. (b) Large hepatic masses. (c) Bilateral homogeneous testicular uptake. (d) Same level as (a) after treatment, shows complete regression of the interatrial mass and partial involution of the muscular involvement.

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

Case 4 (figures 9 and 10)

- Epidemiologic data: 79 year old female.
- Symptoms: epigastric pain, uncompensated high blood pressure, renal failure and heart failure.
- Diagnosis: LBC NHL, stage IV , IPI 4
- Treatment: Bendamustine - Rituximab

1. CECT (figures 9. b and 10. d, e, f) shows bilateral infiltrating renal tumors with larger extension in the left kidney, spreading towards the hilum and enveloping the renal vein, which is permeable. Pleural and pericardial effusion is present. There is also a noticeable but non-specific myocardial
thickness involving the left ventricle and ventricular septum. Multiple mediastinal adenopathies and a focal lesion in the left parotid gland were present (not shown on figures).

2. TTE (figure 9. d, e, f) revealed subtle pericardial effusion and a prominent thickening of ventricular septum and left ventricle showing solid hypoechoic nodules, with systolic and diastolic dysfunction and elevation of the filling pressures.

3. Baseline staging PET/CT proved hypermetabolic infiltrating masses affecting the ventricular wall and septum in several segments corresponding to secondary cardiac lymphoma (figures 9.a and 10. a, b) and hypermetabolic bilateral renal masses (figure 10.c). The study also confirmed the increase of FDG uptake in mediastinal lymph nodes and left parotid gland.

Note the importance of PET/CT in this case to differentiate lymphomatous infiltration of the myocardium from other causes of myocardium thickening going with heart failure such us hypertrophic myocardiopathy.

No response to treatment studies were found as the patient died due to complications of a hip fracture in the following months.
Fig. 9: 79 year old female, LBC NHL stage IV. (b) Myocardial irregular thickening involving the left ventricle and ventricular septum in several segments noticeable in CECT. (c, d, e) TTE show hypoechoic masses within the ventricular myocardium with major compromise of the septum. (a) Fused PET/CT axial image at the same level as (b) demonstrates hypermetabolic infiltrative masses along the septum and ventricular wall.

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES
Fig. 10: Myocardial (a, b) and renal (c) hypermetabolic infiltrative masses PET/CT axial images consistent with the findings in CECT (d, e, f).

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

Case 5 (figure 11)

- Epidemiologic data: 48 year old male.
• Symptoms: multiple cervical bumps, cough and chest pain.
• Diagnosis: LBC NHL, stage II E, IPI 0 (low risk).
• Treatment: 6 courses of R-CHOP.

PET/CT shows cervical and mediastinal nodal lymphoma and contiguous spreading through the aorto-pulmonary window to the left atrium and main pulmonary artery.

Fig. 11: Fused PET/CT images depict bilateral cervical adenopathies (d) which extend caudally towards the mediastinum. The mass in the aortopulmonary space infiltrates the main pulmonary artery (a, e, f) and the left atrium (a, b, c).

References: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

Cases 6 and 7 (figures 12 and 13)

LBC NHL with mediastinal bulky lymphoma. There is a contiguous infiltration of the pericardium, great vessels and cardiac walls.
**Fig. 12**: 35 year old male, LBC NHL stage IV, IPI 4. (a, b, c, d) Fused PET/CT images show mediastinal mass with infiltration of the visceral and parietal pericardium and pericardial effusion. See the involvement of the epicardial fat, the right ventricular wall and mediastinal fat surrounding the great vessels. It also extends to the mediastinal pleura. (e) CECT at the same level as (c).

**References**: HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES
**Fig. 13:** 33 year old female, LBC NHL stage IVB, IPI 2. Fused PET/CT images show a mass within the anterior mediastinum infiltrating the pericardium, superior vena cava, aortic arch and pulmonary artery.

**References:** HOSPITAL UNIVERSITARIO SON ESPASES - Palma de Mallorca/ES

**Images for this section:**
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Conclusion

Integration of PET/CT into lymphoma staging and monitoring facilitates the detection of cardiac involvement and the assessment of response to therapy. In our series, percentage of secondary cardiac lymphoma detected by this method is nearly 8% of patients with diffuse NHL, greater than that observed in baseline studies using CECT as unique image technique. Most common appearance of cardiac lymphoma was hematogenous spreading into the interatrial septum followed by infiltration of cardiac structures by contiguous lymphoma of the mediastinum. The main value of PET/CT in evaluating extranodal lymphoma is the detection of malignant lesions due to its high hypermetabolism, even the smaller ones and those located in the ventricular myocardium. It is useful in cases with large intracavitary masses because it enables us to differentiate neoplastic masses from thrombi.

Recognizing heart involvement by lymphoma in PET/CT could modify the disease management and thereby, prevent cardiac complications by chemotherapy (e.g., cardiac rupture due to rapid mass debulking).

Personal information

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