Unilateral Primary Adrenal Lymphoma: Uncommon Presentation of a Rare Disease Evaluated Using ^{18}F-fluorodeoxyglucose Positron Emission Tomography/Computed Tomography

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Abstract

Primary adrenal lymphoma (PAL) is a relatively rare disease entity with only fewer than 200 cases reported till date. PAL frequently presents with bilateral adrenal involvement and shows male preponderance. We here present a case of PAL in a 65-year-old female with a relatively uncommon unilateral adrenal involvement. The present case depicts that ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography had decisive role in the treatment management of this patient suggesting its potential utility in the management of this rare disease.

Keywords: ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography, diffuse large B-cell lymphoma, primary adrenal lymphoma, primary extranodal non-Hodgkin lymphoma

Introduction

Primary extranodal non-Hodgkin lymphomas (PE-NHL) have an incidence of ~25%–40%, varying with geography. Although no lone definition is universally accepted, the most wisely observed description defines it as the presentation of lymphoma at extranodal site with no/minor-associated nodal involvement, where bulk of the disease is situated at the extranodal site.[1,2] Primary lymphomatous involvement of the adrenal glands (primary adrenal lymphoma [PAL]) is very rare and accounts for <1% of the PE-NHL cases. Most often PAL presents with bilateral involvement (~75% of the cases) of the adrenal glands as large masses and has a male preponderance.[3] ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (F-FDG PET/CT) is an established imaging modality which is widely used in initial staging, treatment response, and recurrence evaluation of lymphomas.[4] We here report a case of PAL with an uncommon presentation as a unilateral mass in an elderly female who was evaluated using ^{18}F-FDG PET/CT during the disease management.

Case Report

A 65-year-old female presented with pain and heaviness in the right flank for the past 2 months. Examination was unremarkable except for mild right hypochondrial

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tenderness. Evaluation with contrast-enhanced computed tomography revealed a large heterogeneously enhancing soft-tissue mass in the right suprarenal area. A whole-body ^{18}F-FDG PET/CT was performed for further characterization, which revealed intense FDG uptake (maximum standardized uptake value [SUVmax] 20.0; ~9.8 cm × 7.0 cm × 7.8 cm) in the soft-tissue mass involving the right adrenal gland [Figure 1a and b]. A few intensely FDG avid (SUVmax 16.7) enlarged retroperitoneal (largest measuring ~3.6 cm × 2.4 cm), and cervical lymph nodes are also noted [Figure 1c]. Unilateral presentation, intense FDG avidity, and associated lymphadenopathy suggested as possibility of adrenocortical carcinoma versus lymphomatous involvement. Fine needle aspiration cytology from the right adrenal gland mass revealed lymphomatous infiltration, and a core needle biopsy performed subsequently yielded a histopathological diagnosis of diffuse large B-cell lymphoma (DLBCL) through immunotyping [Figure 2a-d]. Bone marrow aspiration was negative for lymphomatous infiltration, and the patient was staged as Ann Arbor Stage IIIE. The patient then received four cycles of R-CHOP regimen, and interim ^{18}F-FDG PET/CT revealed faint FDG avidity (SUVmax 2 vs. 20 previously) of the right adrenal mass lesion and resolution of FDG avidity of the retroperitoneal and cervical lymphadenopathy suggesting a favorable response to therapy [Figure 1d-f]. The patient then received two more cycles of R-CHOP. ^{18}F-FDG PET/CT done at the end of six cycles revealed mild persistent FDG avidity in the right adrenal mass and appearance of renal lesions (SUVmax 18.3) and paracaval lymph nodes suggesting disease progression [Figure 1g-i]. Rapid cytopenia developed in the patient following chemotherapy, and the patient was changed to alternate chemotherapy regimen (R-ICE). Further deterioration in thrombocytopenia was noted in the patient after the first cycle, which lead to discontinuation of therapy and was followed by symptomatic treatment. Currently, the patient is clinically stable and started on oral chemotherapy regimen containing endoxan, etoposide, and procarbazine.

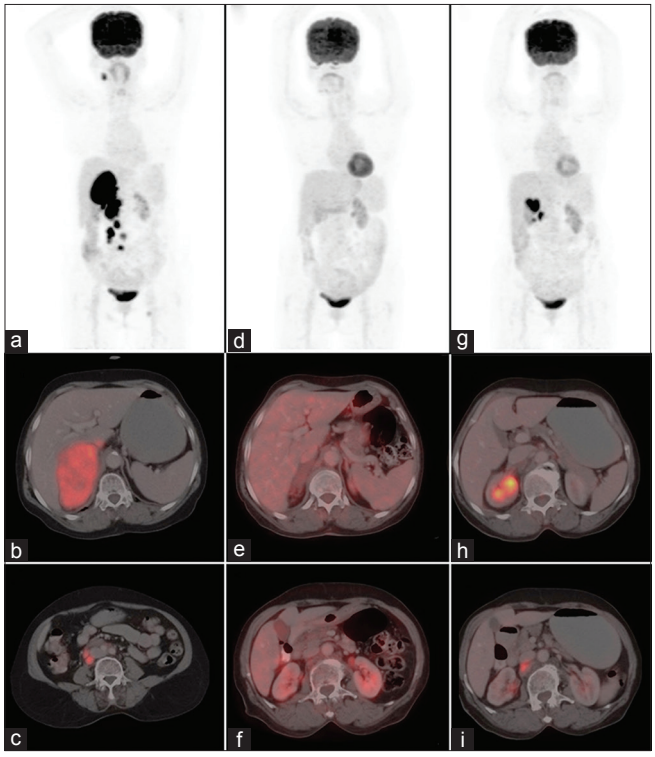

Figure 1: (a) Maximal intensity projection of the staging ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography showing areas of intense abnormal tracer uptake in the right suprarenal location and along the midline; (b and c) Transaxial fused positron emission tomography/computed tomography images localized the intense tracer uptake to a large suprarenal mass and to a few enlarged retroperitoneal lymph nodes; (d-f) maximal intensity projection and fused positron emission tomography/computed tomography images of interim positron emission tomography/computed tomography (following four cycles of chemotherapy) showing significant resolution in size and fluorodeoxyglucose avidity of the right suprarenal mass and retroperitoneal lymph nodes. (g-i) End of treatment positron emission tomography/computed tomography images (after two more cycles of chemotherapy) revealing disease relapse in the form of a right renal lesion and a few retroperitoneal lymph nodes

Discussion

Normally adrenal glands are devoid of lymphatic tissue and hence it is often believed that primary lymphomatous involvement originates in the setting of a precedent autoimmune adrenalitis.[5] There is paucity of literature evaluating large number of PAL, likely attributable to its rarity. In a recent didactic review of 187 cases by Rashidi and Fisher, the authors have systematically

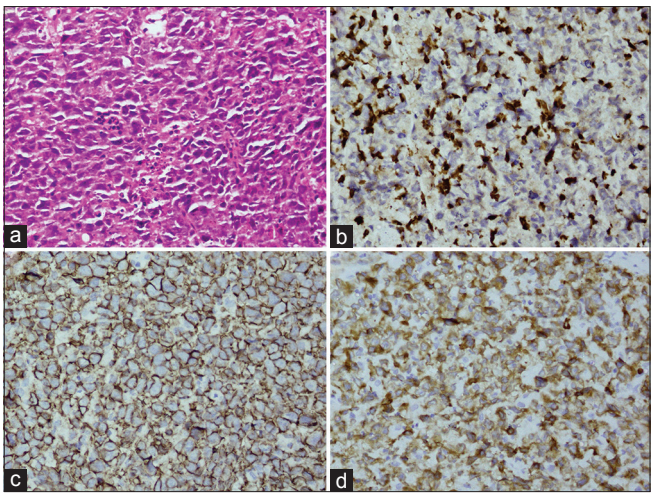

Figure 2: Photomicrographs showing (a) sheets of atypical lymphoid cells with large nuclei and moderate cytoplasm (H and E, ×400); (b) membrane positivity of tumor cells for CD20; (c) atypical lymphoid cells showing positivity for CD79a; (d) CD3 immunostaining highlighting reactive cells (b-d: Immunoperoxidase, ×400) confirming a diagnosis of diffuse large B-cell lymphoma

summarized the available cases of PAL in literature. They showed that PAL has a preferential bilateral involvement with DLBCL (~78% of the cases) as the most common histology. B-symptoms, abdominal pain, and fatigue were the most common symptoms at presentation. In their series, the authors also noted a high incidence of adrenal insufficiency (~61%), more commonly associated with bilateral adrenal gland involvement.[3]

Conventionally, PE-NHLs are claimed to demonstrate poorer prognosis as compared to nodal lymphomas with a higher relapse rate. Former cases of PAL treated with CHOP/CHOP-like regimens reported shorter survival rates. Kumar *et al*. in their study reported a mean overall survival of only 15.3 months (3.6 vs. 34 months in those with no response and that showed disease remission, respectively).[6] In a single largest study in 31 patients by Kim *et al*., the authors incorporated rituximab to the chemotherapy regimen. They reported higher survival rates as compared to the previously reported literature.[7]

DLBCL, an aggressive histological subtype, is usually highly metabolically active and being the most common of subtype noted in PAL, facilitates imaging with metabolic imaging using ^{18}F-FDG PET/CT. The established role of ^{18}F-FDG PET/CT in the management of metabolically active lymphomas permits its translation into evaluating of this rare entity.[4,8] The present case depicts the utility of ^{18}F-FDG PET/CT in assessment of disease response and also early detection of progression/relapse.

Furthermore, in the present case, several features including unilateral involvement, female gender, and the absence of several common associations were against an etiology of PAL. Thus, this case illustrates an uncommon presentation of PAL and suggests the need to consider it as a differential diagnosis during evaluation of adrenal masses.

Conclusion

^{18}F-FDG PET/CT is potential modality in the evaluation of patients with PAL for initial staging, response evaluation, and early detection of disease progression and thus can aid in therapeutic decision-making.

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Conflicts of interest
There are no conflicts of intrest.

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