Case Report

Unilateral primary adrenal natural killer/T-cell lymphoma: Role of fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography for staging and interim response assessment

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ABSTRACT

Primary adrenal lymphoma (PAL) is a rare malignancy often involving bilateral adrenal glands. Diffuse large B-cell is the most common histological type. Unilateral presentation and T-cell/natural killer (T/NK) cell histological type is rarer. We report fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography scan findings in a case of unilateral T/NK cell PAL performed for staging and interim treatment response assessment.

Keywords: Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography, primary adrenal lymphoma, T-cell/natural killer cell lymphoma

INTRODUCTION

Extranodal involvement is seen in approximately 25% cases of nonHodgkin's lymphoma (NHL). Primary adrenal lymphoma (PAL) is rare with fewer than 200 cases reported in English literature. The majority of the patients with PAL present with bilateral adrenal masses and diffuse large B-cell (DLBCL) type is the most common type on histology. Unilateral presentation and T-cell/natural killer (T/NK) cell histological type is rarer.[1-3] Due to the rarity of the disease, there is a paucity of literature on the role of fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (F-18 FDG PET/CT) scan in PAL. We report F-18 FDG PET/CT scan findings in a case of unilateral T/NK cell PAL performed for staging and interim treatment response assessment.

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CASE REPORT

A 41-year-old gentleman with no co-morbidities presented with the progressive left hypochondriac pain of 1-month duration. Karnofsky performance score was 90%, and Eastern Cooperative Oncology Group score was 0. Contrast enhanced CT revealed 8 cm × 7 cm sized enhancing left suprarenal mass. The clinical and biochemical parameters ruled out adrenal insufficiency and pheochromocytoma. The serum lactate dehydrogenase (LDH) level was raised. CT guided biopsy with immunohistochemistry revealed tumor cells positive for leukocyte common antigen, CD3, and weakly positive for CD56; while negative for CD20, CD4, and CD8 suggestive of high-grade NHL of T/NK cell immunophenotype [Figure 1]. Bone marrow biopsy was unremarkable. Staging F-18 FDG PET-CT scan revealed increased metabolic activity in the left adrenal mass. No focus of hypermetabolism was noted elsewhere in the body suggestive of

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Stage I Aex (left adrenal) [Figure 2]. The patient was started on "SMILE" (methotrexate, dexamethasone, leucovorin, ifosfamide, etoposide, and L-asparaginase) chemotherapy regimen. Interim FDG PET-CT for response assessment post four cycles of chemotherapy revealed complete metabolic and morphological regression of the primary left adrenal mass. No new lesions were noted suggestive of complete metabolic and morphological response to therapy.

DISCUSSION

PAL is defined as the presence of adrenal lymphoma without any evidence of nodal involvement or leukemia. Although secondary adrenal involvement in NHL is seen in approximately 25% of cases, PAL is rare comprising <1% of NHL and 3% of extranodal NHL with <200 cases reported in English literature. Unilateral adrenal lymphoma, as seen in our patient, is even rarer.[4-6]

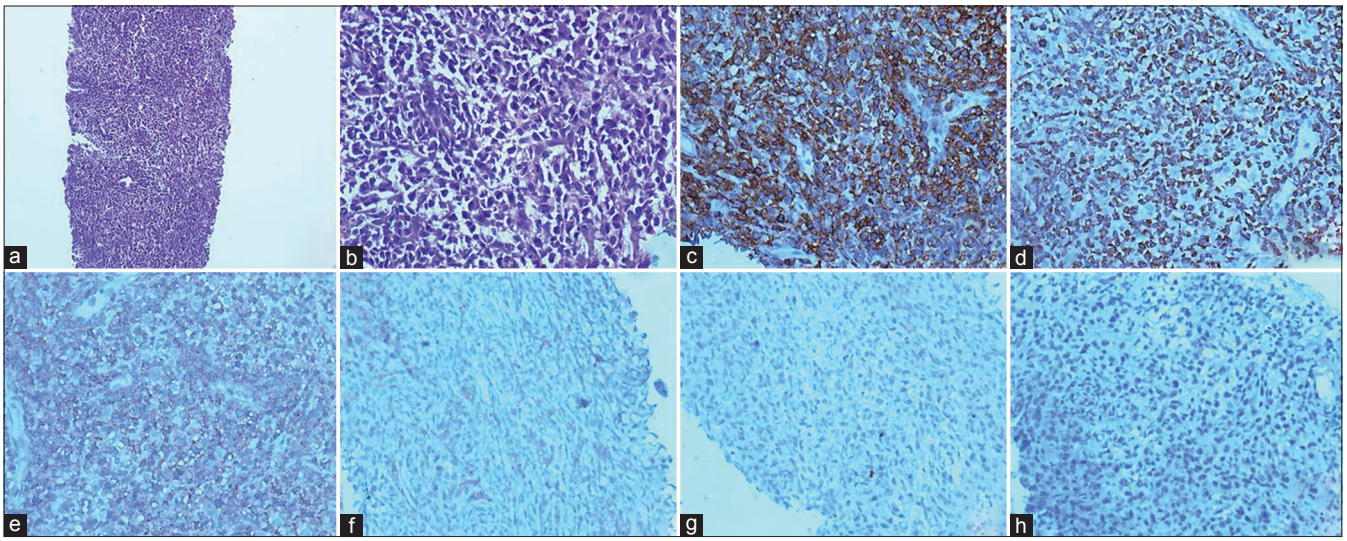

Figure 1: Biopsy of the left adrenal mass: (a and b) Photomicrographs showing features of nonHodgkin's lymphoma with diffuse architecture composed of intermediate to large atypical lymphoid cells (H and E, ×4 and ×40 respectively). (c) Immunohistochemistry is positive for leukocyte common antigen (×40). (d) CD-3 (×40). (e) CD-56 (×40). Immunohistochemistry was negative for (f) CD-20 (×40). (g) CD-4 (×40). (h) CD-8 (×40)

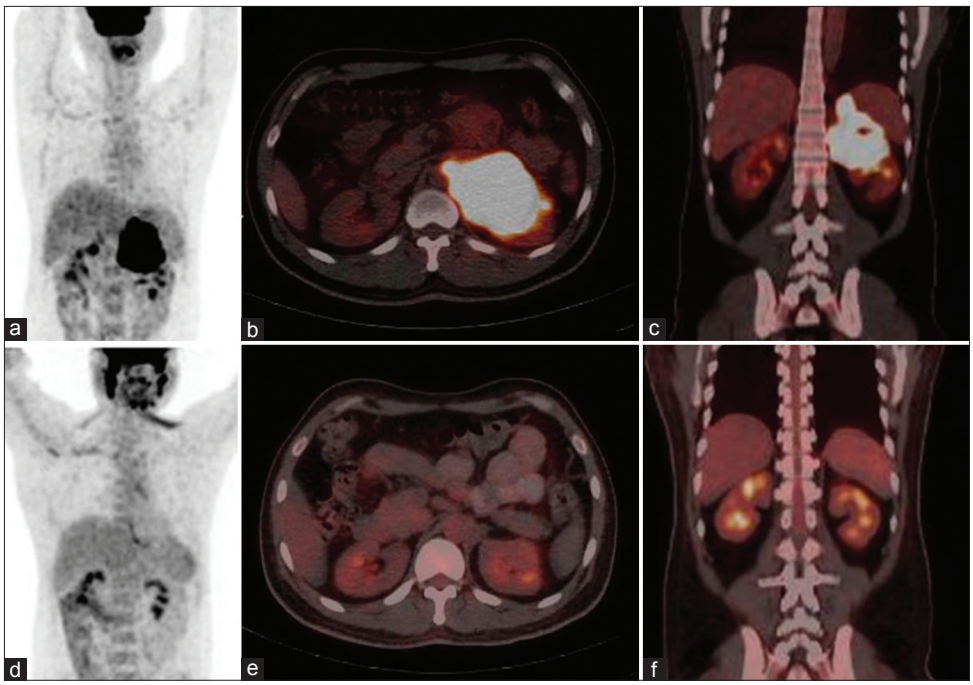

Figure 2: Increased fluorodeoxyglucose uptake was noted in the left suprarenal gland on maximum intensity projected: (a). Axial positron emission tomography/computed tomography. (b) Coronal positron emission tomography/computed tomography. (c) Images of the staging positron emission tomography/computed tomography scan in case of biopsy proven adrenal lymphoma. Positron emission tomography/computed tomography scan done post four cycles of "SMILE" chemotherapy regimen revealed complete metabolic and morphological response (d-f)

PAL is seen more commonly in the elderly age group with male preponderance. The mean age of diagnosis is 68 years. Most of the patients present with nonspecific symptoms such as abdominal pain, fever, and weight loss. In 60% cases, bilateral adrenals are involved and can be associated with adrenal failure in 66% patients. Advanced age at diagnosis, large tumor size, increased LDH level, bilateral adrenal involvement (regarded as Stage IV disease), and adrenal insufficiency at the time of presentation are poor prognostic factors.[2]

The pathogenesis of PAL is still not clear. Since adrenal gland is devoid of lymphoid or hematopoietic tissue, PAL is believed to originate from differentiation of primitive totipotent mesenchymal cells. It is also believed to arise from preexisting autoimmune adrenalitis with lymphocyte infiltration. However, none of these theories has been conclusively proven due to the rarity of the disease.[7,8]

The treatment of PAL depends on the histological subtype. Histologically, the most common type of PAL is DLBCL.[1] There are only 5–6 cases of T/NK-cell type PAL reported in the literature so far, most of them with bilateral adrenal involvement. Unilateral presentation of PAL with T/NK cell immunophenotype makes our case unique.

Conventionally, CT scan is used as a first imaging modality for characterization of adrenal lesions. T2-weighted magnetic resonance imaging with chemical shift sequences is also often used to differentiate benign from malignant adrenal masses.[9]

F-18 FDG PET/CT scan has an established role in staging, restaging, interim response assessment, and prognostication of lymphomas. Various studies have shown higher accuracy of F-18 FDG PET/CT scan in differentiating malignant and benign adrenal masses. Yun *et al.*[10] have shown that FDG PET has a 100% sensitivity, 94% specificity, and 96% accuracy for characterization of adrenal lesions. However due to the rarity of the disease, there is a paucity of literature on the role of FDG PET/CT scan in PAL.

In our case, baseline FDG PET/CT showed intense uptake in the left adrenal mass with no other nodal or extranodal disease detected elsewhere in the body. Hence, in correlation with the biopsy findings, whole body FDG PET/CT played a paramount role in differentiating primary versus secondary involvement of adrenal gland. A study by Khong *et al.* assessing the role of mid-treatment FDG PET/CT for early response assessment of SMILE therapy in NK/T-cell lymphoma demonstrated that Deauville score on mid and end treatment FDG PET/CT is the only significant independent predictor of both overall survival and progression free survival. To the best of our knowledge, this is the first case reported in the literature where a complete metabolic and morphological response was seen on interim FDG PET/CT in a case of unilateral PAL of T/NK-cell type treated with SMILE regimen.[11-16] Therefore, our case further reinforces the valuable role of FDG PET/CT for diagnosis, staging, and treatment response evaluation in PAL.

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Conflicts of interest

There are no conflicts of interest.

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