

Primary Non Hodgkin's Lymphoma of Left Adrenal Gland – A Rare Presentation

ANIL KHURANA¹, PARAMJEET KAUR², ASHOK K CHAUHAN³,
SANT PARKASH KATARIA⁴, NUPUR BANSAL⁵

ABSTRACT

Primary adrenal lymphoma is rare and constitutes for 3% of extranodal lymphoma cases. Approximately 70% of patients present with bilateral disease and have adrenal insufficiency. Prognosis of primary adrenal lymphoma (PAL) is poor, most of patient die within one year of diagnosis. Moreover, there are no standard treatment protocols on such cases. Patients are generally treated with regimens similar to other nonhodgkin lymphoma which includes surgery, combination chemotherapy and or radiotherapy. We are presenting a successfully treated case of primary diffuse large B cell non Hodgkin lymphoma of adrenal gland in a 57-year-old patient. The patient had unilateral adrenal involvement (left adrenal gland), without adrenal insufficiency and normal Serum lactate dehydrogenase level. The patient was treated with left adrenalectomy followed by combination chemotherapy. Two years after diagnosis and treatment the patient is disease free on clinical and imaging studies.

CASE REPORT

A 57-year-old male patient presented with history of recurrent on and off fever of three months duration. There was no history of pain, anorexia, weight loss, nausea and vomiting. Patients hematological, kidney function test and liver function test were within normal limit. There was no lymphadenopathy or organomegaly. Chest X-ray of the patient was normal. USG whole abdomen showed a renal mass. Contrast Enhanced Computed Tomography (CECT) abdomen and pelvis was done which showed 8.9 x 6.3 cm mass in left suprarenal region [Table/Fig-1]. The patient underwent left adrenalectomy. Cut surface showed a Grey- White solid tumour with a lobulations, yellow areas and necrosis. The tumour was occupied complete mass. Microscopic examinations showed a tumour composed of neoplastic cells arranged in nodules and diffuse solid sheets separated by thick fibrous strands. The neoplastic cells had pleomorphic nuclei with prominent nucleoli and eosinophilic cytoplasm. Mitotic activity was high ~ 20 / 10 hpf, large tract of necrosis and apoptoses were visible. The tumour involved the capsule.

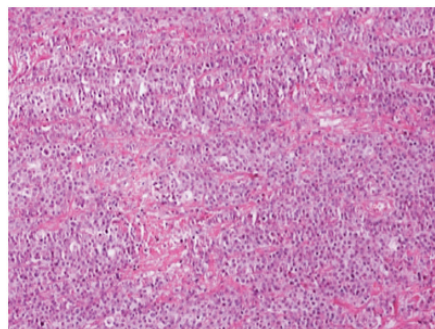


[Table/Fig-1]: Preoperative CECT abdomen and pelvis showing left suprarenal mass

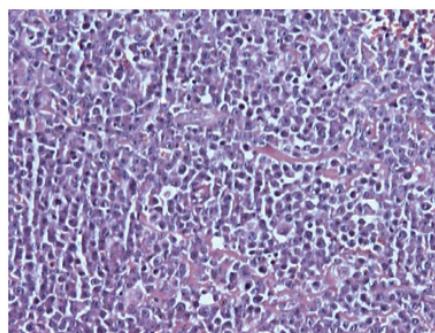
On immunohistochemistry the tumour cells are positive for LCA and negative for CK, Melan A, Inhibin and calretinin. In addition cells are reactive for CD20, lack expression of CD 3 [Table/Fig-2a-2f]. The patient was diagnosed to having Diffuse Large 'B' cell Lymphoma of left adrenal gland. Serum Plasma adrenocorticotrophic hormone level and cortisol level of patient were within normal limits. Bone marrow examination and bone marrow biopsy were negative for tumour involvement.

Keywords: Adrenal tumour, Chemotherapy, Extranodal lymphoma

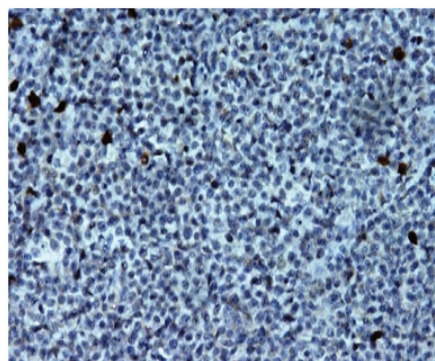
Positron Emission Tomography (PET) scan confirmed the diagnosis of *Primary* Diffuse Large B cell Lymphoma of Left Adrenal gland



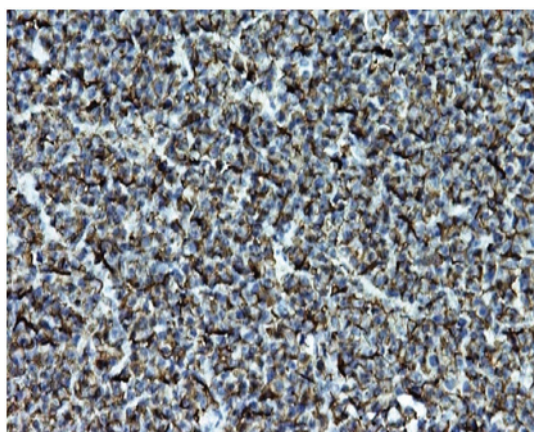
[Table/Fig-2a]: Showing tumour cells arranged in nodule & nests separated by thick fibrous septa. (H&E 100X)



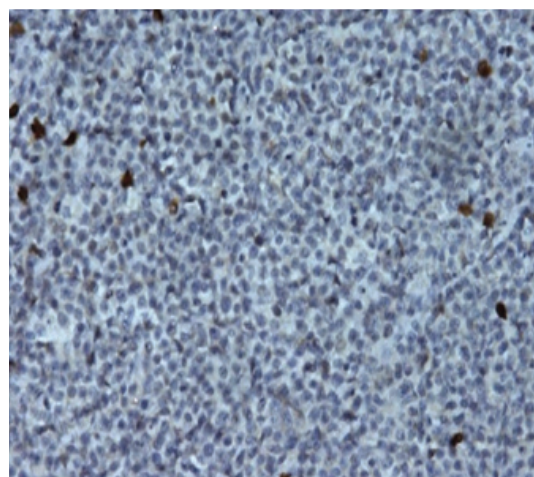
[Table/Fig-2b]: Uniform population of lymphoid cells (H&E 400X)



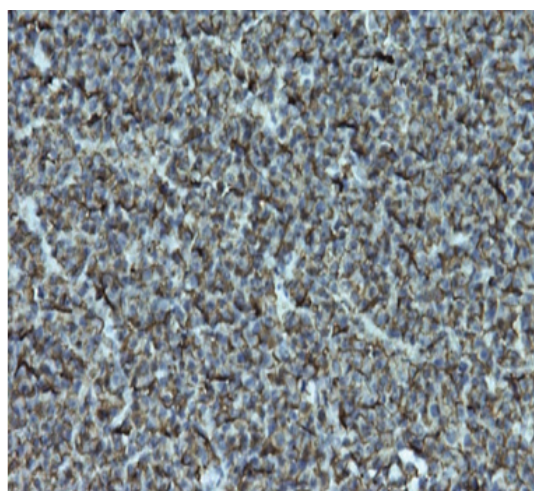
[Table/Fig-2c]: Negativity for cytokeratin (Immunoperoxidase Staining 100X)



[Table/Fig-2d]: Showed Tumours cells positive for Leucocyte Common Antigen (LCA) (Immunoperoxidase Staining 400X)



[Table/Fig-2e]: Tumour cells lack expression of CD 3



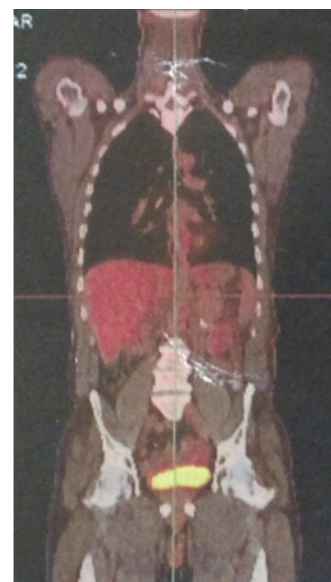
[Table/Fig-2f]: Tumour cells positive for CD20

[Table/Fig-3]. Then the patient was administered combination chemotherapy with Injection Rituximab 500 mg, Injection Cyclophosphamide 1 gm, Injection Vincristine 2 mg and Tablet Prednisolone 100 mg orally for five days (R-CHOP) three weekly for six cycles.

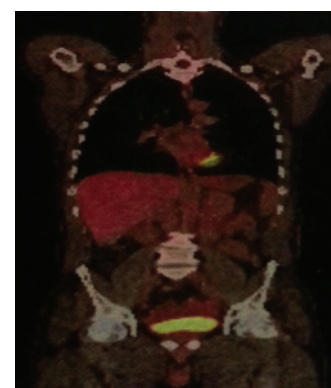
Follow up PET scan study did not show any evidence of local recurrence [Table/Fig-4].

DISCUSSION

Primary adrenal lymphoma is defined as malignant neoplastic proliferation of the lymphoid cells exclusively in the adrenal glands. Adrenal involvement of disseminated lymphoma is common and reported up to 24% of cases [1]. However, primary adrenal non Hodgkin lymphoma is rare and constitutes less than 1% all NHL and 3% of extranodal lymphoma [2,3]. Approximately 120 cases



[Table/Fig-3]: Positron Emission Tomography scan - postoperatively showing no evidence of disease and no dissemination from any other sites. Suggestive of Primary Non Hodgkin lymphoma of left adrenal gland



[Table/Fig-4]: Positron Emission Tomography scan at last follow up (one year)- No evidence of disease

of primary adrenal lymphoma as case reports or case series have been reported in English literature [3]. Bilateral involvement of adrenal glands is reported in 70% of cases [4]. This tumour affects predominantly elderly and males with male to female ratio (2:1). Histopathological examination and immunohistochemistry is standard for confirmation of diagnosis. The diffuse large B – cell lymphoma histology is common and reported in 70% of cases.

The present case is a diffuse large cell B – Cell lymphoma of adrenal in 57-year-old male patient which is in concurrence with cases reported in literature [5,6]. There are no definitive treatment protocols for these tumours. Such tumours are treated as standard treatment of non Hodgkin lymphomas. Treatment modality includes surgery, multi agent combination chemotherapy, radiotherapy and corticosteroid replacement therapy.

The R-CHOP chemotherapy regimens are a standard treatment of Non Hodgkin lymphoma. The patient completed six courses of chemotherapy and did not show any evidence of recurrence, relapse during treatment and after two years of treatment on clinical examinations and on follow up PET scan imaging studies.

Prognosis of Primary Adrenal Non Hodgkin Lymphoma is dismal and these tumours are considered lethal. Most of the patients reported in literature showed a median survival of 12.5 weeks despite aggressive chemotherapy [7]. Whereas, only infinitesimal numbers of cases have been reported so far of primary NHL adrenal that showed complete or partial remissions with a longer mean duration of survival [8,9].

Prognostic factors includes age, adrenal insufficiency, elevated serum Lactate dehydrogenase, tumour size may have significant impact on treatment outcome and survival. Adrenal insufficiency

usually occurs in bilateral involvement of adrenal glands [10]. Plasma adrenocorticotrophic hormone level and cortisol level should be within normal limits to rule out adrenal insufficiency.

As in present case the patient had unilateral adrenal gland involvement, normal Serum Lactate Dehydrogenase level and no adrenal insufficiency that factors might be considered for complete response and relapse free survival. Moreover, the present case disease was operable and patient underwent radical surgery followed by R-CHOP chemotherapy.

Aziz et al., reported two cases of primary adrenal lymphoma. One with unilateral adrenal involvement had a normal adrenal function and other with bilateral involvement had adrenal insufficiency. The first case with unilateral adrenal involvement has shown dramatic subjective and radiological response to chemotherapy (gained body weight and had marked regression in the size of affected adrenal gland on CECT abdomen), while as second patient died after two cycles of chemotherapy [11].

Moreover, longer duration of survival and complete remission was reported in studies when treated with R-CHOP [3,12-14].

Kanwar et al., in their study reported two cases of primary adrenal lymphoma. In first case had bilateral adrenal involvement with adrenal insufficiency managed by combination chemotherapy with Rituximab – CHOP was doing well till last follow up [12].

Kim et al., reported a case of bilateral primary adrenal lymphoma received only R-CHOP chemotherapy, without surgery, radiation or high-dose therapy. There is no evidence of recurrence after 12 months [13].

CONCLUSION

Primary Lymphoma of adrenal gland is a rare entity with poor prognosis. Most cases reported are bilateral with adrenal insufficiency therefore resulted in poor prognosis. However, the patient may achieve complete response with unilateral and localized involvement of adrenal gland without nodal involvement and without adrenal

insufficiency. Early diagnosis and aggressive multimodality treatment strategies including surgery and combination chemotherapy may have impact on outcome and long term survival.

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PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Radiotherapy, Regional Cancer Centre, Pt. BD Sharma. PGIMS, Rohtak, India.
2. Associate Professor, Department of Radiotherapy, Regional Cancer Centre, Pt. BD Sharma. PGIMS, Rohtak, India.
3. Senior Professor, Department of Radiotherapy, Regional Cancer Centre, Pt. BD Sharma. PGIMS, Rohtak, India.
4. Professor, Department of Pathology, Pt. BD Sharma. PGIMS, Rohtak, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Anil Khurana,
Senior Resident, Department of Radiotherapy, PGIMS, Rohtak, Haryana, India.
E-mail : drparamjitkaur@rediffmail.com

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