



A RARE CASE OF NON HODGKIN'S LYMPHOMA OF ADRENAL GLAND

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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 64-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 debulking surgery followed by combination chemotherapy.

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INTRODUCTION

The involvement of certain organs such as the adrenal gland is rare in non-Hodgkin's lymphoma (NHL). Although lymphomas can involve virtually all extranodal organs, different organs show different frequency of involvement. Adrenal gland involvement is usually bilateral. We report that a rare case of unilateral adrenal involvement of diffuse large B-cell lymphoma presenting with abdominal pain.

Case Report

A 64 year old male patient, resident of kadegaon, Sangali, came to Krishna hospital, karad, with chief complains of lump in abdomen since 10 years and pain in abdomen since 2 months. Patient noticed lump in the left side of abdomen which was insidious in onset, progressive in nature, which was associated with pain of mild to moderate intensity and pain was increased on aggressive work, having food and decreased on taking rest. Patient is a known case of hypertension and asthma since 10 years and on medications for the same. On general examination, vitals were normal, pallor present, no icterus, cyanosis, clubbing, oedema, lymphadenopathy.

Per abdominal examination : lump palpable in left hypochondrium region of size 7 x 5 cm irregular in shape, firm in consistency, non-tender, moves with respiration, mobile side to side and on head raising test it was intra-abdominal swelling, no organomegaly, bowel sounds present.

On investigation:

Blood reports: Hb 8.8 g/dl increased to 9.7 g/dl after blood transfusion. TLC 6400/ul, Platelet count 2.10 lakhs/cumm, Renal function test and serum electrolytes all normal. HIV.HbsAg, HCV all non reactive.

Chest Xray: Basal segments of both the lung fields show increased reticulonodular areas of opacities.

Usg abdomen and pelvis(dated 4/2/2019): An ill-defined heterogeneously hyperechoic mass with anechoic areas within noted in the region of body and tail of pancreas. The lesion is seen extending upto splenic hilum causing mass effect on left kidney displacing it inferolaterally. The lesion also appears to be encasing coeliac trunk. The visualized part of head of pancreas appears normal however body and tail could not be assessed separately from the lesion. The lesion is likely originating from pancreas. The lesion shows vascularity on colour Doppler. An anechoic oval structure noted in the region of tail of pancreas is likely to be a necrotic lymph node.

CECT abdomen and pelvis(dated 6/2/2019): An ill defined mildly enhancing soft tissue attenuation mass lesion with few non enhancing regions within in retro peritoneum on left side, not seen separately from left adrenal gland, invading the upper pole of left kidney and encasing the vessels as described-neoplastic etiology.

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24 hours Urinary V.M.A levels: 9.7 mg/day (normal range- 2 to 14 mg/day) Debulking surgery was done under general anaesthesia on 15/2/2019 and specimen sent for histopathology.

HPR report dated 26/2/2019(B/1563/19): High grade malignancy suggestive of high grade Carcinoma- excised specimen from upper pole of left Kidney. Suggested differential diagnosis:

1. Adrenocortical tumour
2. Other poorly differentiated carcinoma.

Immunohistochemistry (dated 5/3/2019): High grade B cell non Hodgkin's Lymphoma.

Bone marrow study: Micronormoblasticaemia with eosinophilia with reactive plasmocytosis.

Intraoperative Photos



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. However, primary adrenal non Hodgkin lymphoma is rare and constitutes less than 1% all NHL and 3% of extranodal lymphoma. Approximately 120 cases of primary adrenal lymphoma as case reports or case series have been reported in English literature . Bilateral involvement of adrenal glands is reported in 70% of cases . 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 64-year-old male patient which is in concurrence with cases reported in literature . 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. 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.

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. Plasma adrenocorticotrophic hormone level and cortisol level should be within normal limits to rule out adrenal insufficiency.

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