ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
Index Copernicus Value: 49. 23
PubMed - National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 4, Issue – 3, May - 2019, Page No. : 44 – 48

Unilateral Primary Adrenal Diffuse Large B- Cell Lymphoma with Metastatic Nodule on Spleen – A Rare Case Report

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Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Primary haematolymphoid tumours involving the adrenal gland are infrequent and are mainly lymphomas. Primary adrenal lymphoma (PAL) is uncommon in clinical settings, and is considered as a rare high grade malignant lymphoma with poor prognosis. Patients with unilateral mass with normal adrenal functioning are extremely rare. Patient may show secondary involvement of lymph nodes, bones, spleen, lungs, gastrointestinal tract, central nervous system and other endocrine organs.

We report a case of a 49year post-menopausal female presented with left lumbar quadrant abdominal pain. Local examination showed firm palpable lump on deep palpation. Haematological and biochemical profile was within normal limits. Abdominal ultrasonography and abdominopelvic CT showed an enlarged left adrenal mass, likely of a neoplastic etiology with abdominal pre and para-aortic lymphadenopathy. Histopathological examination and immunohistochemistry confirmed the diagnosis of Diffuse Large B -Cell Lymphoma of the left adrenal gland with secondary involvement of splenic nodules. The present study highlights rare occurrence of primary adrenal lymphoma as the unilateral adrenal mass with secondary involvement of splenic nodules.

Keywords: primary adrenal lymphoma, diffuse B cell lymphoma, immunohistochemistry, adrenal cortical carcinoma, spleen, metastasis

Introduction

Non-Hodgkins Lymphoma (NHL) typically arises from the lymphoid tisues, although nearly 25-40%of the cases arise in extra-nodal sites (1).

Primary adrenal lymphoma (PAL) is adrenal lymphoma arising within and confined to adrenal glands, without nodal involvement or a leukemia(2)

Primary adrenal lymphoma (PAL) is very rare and accounting for less than 1 % of all Non-Hodgkins Lymphoma cases(3) with commonest being Diffuse Large B Cell Lymphoma(4) . Most of the cases show bilateral adrenal involvement and common symptoms of fever, weight loss, abdominal /lumbar pain and symptoms of adrenal insufficiency(5,6)

Several prognostic factors have been considered in the management of primary adrenal lymphoma, such as age (more than 60 years), presentation with adrenal insufficiency, tumour size (more than 10 cm), bilateral involvement LDH and β2 microglobulin levels and involvement of other organs.

Being highly aggressive neoplasm, primary adrenal lymphoma with secondary extra nodal involvement is associated with poor prognosis and 1 year survival rate of 17.5% (7)

Case report

A 49 year old post menopausal female was referred to our hospital with pain in the left lumbar region of the abdomen, radiating to the back since 3 months. The pain was dull aching and gradually progressive in nature. She also complained of fever with chills since 3 days. She also had history of significant weight loss and loss of appetite. There were no similar complaints in the past or history of any medical or surgical illness. On clinical examination, there was no lymphadenopathy or organomegaly.

On per abdomen examination a palpable firm lump was felt in the left lumbar region on deep palpation, which moved with respiration.

Lab investigations were as follows: Sodium-Potassium level of 131 mEq/L (reference range -133-150 mEq/L) and 3.1mEq/L (reference range -3.3-4 mEq/L) respectively. Blood urea and Sr.creatinine of 11mg/dl reference range-14-36mg/dl) and 0.54mg/dl(respectively.

VMA (Vanillyl Mandelic Acid) SPOT – Negative

24 hour urine metanephrine-293µg/24 hrs (normal range -25-312.)

Ultrasound abdomen-pelvis revealed a heterogenous lesion arising from the left suprarenal gland, probably of neoplastic etiology along with abdominal lymphadenopathy.

Abdominal Computed Tomographic (CT)scan revealed an ill-defined,lobulated heterogenous enhancing lesion near the upper pole of the left kidney.The left adrenal gland was not visualised separately. Multiple enlarged heterogenous lymph nodes were seen in the pre and para aortic, aorto-caval and peri -pancreatic region.

Patient underwent exploratory laparotomy for left adrenalectomy with splenectomy. Multiple enlarged lymph nodes were seen near the superior border of the pancreas and near the splenic hilum . An enlarged mass of 12x8cm involving the left adrenal gland was resected and splenectomy was done .The specimen were sent for histopathological evaluation.

Pathologic Findings

A left adrenalectomy specimen weighing approximately 400 gms and measuring about 12.5x10.2x5 cm was received . Gross appearance and cut surface of adrenal mass has been shown in Figure 1and 2. The resected bluish grey spleen measuring 11x8.5 x 4.5cm was received and had smooth external surface with attached 3 small nodules . Largest nodule is seen near hilum measuring 1.5x1x0.5cm as shown in figure 3

Histopathological examination

Revealed sheets & nest of tumour cells arranged in cords, trabeculae and diffuse growth like pattern. (as in figure 4,5)These are large round to oval, hyperchromatic nuclei with prominent nucleoli (as in figure 6) & moderate amount of eosinophilic cytoplasm. There is moderate anisonucleosis & pleomorphism. Mitotic figures (15-20/10hpf) with few pleomorphic to bizzare cells seen. Vascular invasion seen. Areas of hemorrhage and necrosis seen. Adjacent area showed normal adrenal tissue. Histologically, it was reported as poorly differentiated adrenal cortical carcinoma, however Large B-cell lymphoma cannot be ruled out, for which

immunohistochemistry will be necessary Splenectomy sections showed normal splenic tissue, but splenic nodules were secondarily involved by neoplastic cells as shown in figure 7.

Immunohistochemistry evaluation revealed tumour cells expressing leucocyte common antigen LCA, CD 20, CD79a,BCL-2 while negative for Cytokeratin , CD3 and CD 10 with Mib -1 proliferation index of 70 % leading to diagnosis of Non- Hodgkin Lymphoma – Diffuse Large B-cell Lymphoma (DLBCL) phenotype

Discussion

ACC is rare with an annual incidence of less than 0.7 to 1.5 per 1 million and leading to limited prospective studies. [8] Although it can present in any age has bimodal presentation with peaks in 1st (1- 6 yrs) and 4th-5th decade of life. Female predominantly affected with M:F ratio of 2.5:1(9).

Primary adrenal lymphoma is a rare extranodal lymphoma representing only 3% of extranodal lymphomas, with 100 cases been reported in last 40 to 50 years. (4)

An EBV association is observed in B- cell and T cell lymphomas. B cell lymphomas are predominantly seen with DLBCL being commonest. Plasmablastic lymphoma, Burkitt lymphoma and low grade B cell lymphomas are also reported.

In this case report , left adrenal mass was markedly enlarged as compared to associated lymphadenopathy and without presence of leukemia suggesting primary involvement of unilateral adrenal. (9)

Clinical presentation of primary adrenal lymphoma may vary widely. Patients present with fever, weight loss,abdominal pain and symptoms of adrenal insufficiency,while few remains asymptomatic.

Parameswaran R et al stated DLBCL had high incidence of bilateral adrenal involvement and low incidence of extra-adrenal disease at the time of diagnosis and reported a 58 year old Primary Adrenal Diffuse Large B Cell Lymphoma.[4]

Unilateral PAL are generally not been associated with adrenal insufficiency cases zhang et al reported a case of an 80 year old with Primary Unilateral Adrenal Burkitt-Like Large Cell Lymphoma presenting as adrenal insufficiency. [11]

Lei.Z et al presented 2 cases of bilateral adrenal Primary Diffuse Large B Cell Lymphoma without adrenal insufficiency.[12]

Secondary adrenal metastasis is seen in 27 % of adrenal carcinoma cases.(9)

DLBCL consist of diffuse growth pattern of medium to large B lymphoid cells whose nuclei is more than twice that of normal lymphocyte. DLBCL-NOS constitute of approximately 25- 30 % of Adult Non- Hodgkins Lymphoma and express specific antigens of B lymphocytes (CD 20 and CD 79a)

DLBCL NOS has morphological variant such as centroblastic , immunoblastic anaplastic and other rare variants and molecular subtypes of germinal centre B cell subtype and activated B cell subtype.

Genetic profile -Most primary DLBCL of adrenal gland are of activated B cell type with majority expressing BCL 6 gene rearrangement.(9)

Conclusion

Although rare , DLBCL should be considered in the differential diagnosis, even in unilateral adrenal cortical carcinoma cases with or without adrenal insufficiency for early diagnosis and potentially improve survival rates. Histopathological examination and immunohistochemistry been standard for confirmation of diagnosis.

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Legend Figure

Figure 1: Left Adrenalectomy specimen. Figure 1- Gross appearance of an enlarged, well encapsulated, tan grey left adrenal mass with nodular external appearance alond with 3 separately received tissue pieces.

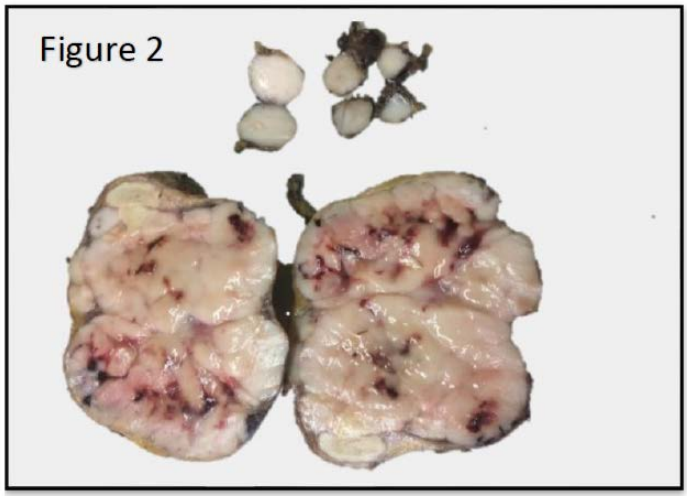


Figure 2- cut section of mass showing fish-flesh appearance with haemorrhagic areas and that of the separately received pieces is homogenous white.

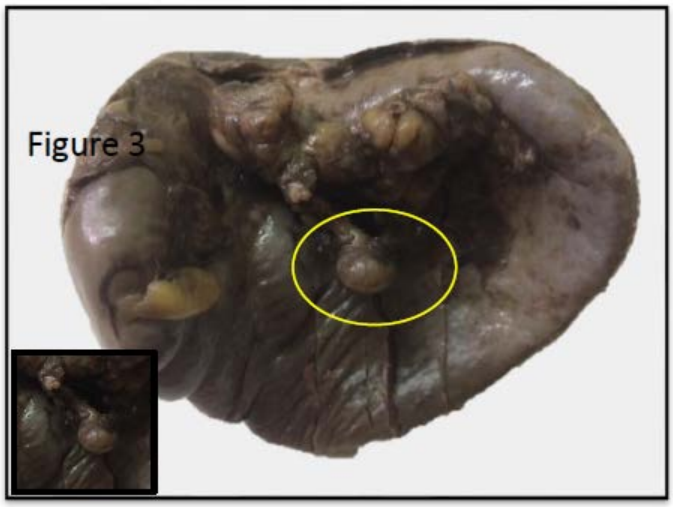


Figure 3 Splenectomy specimen showing largest nodule near hilum

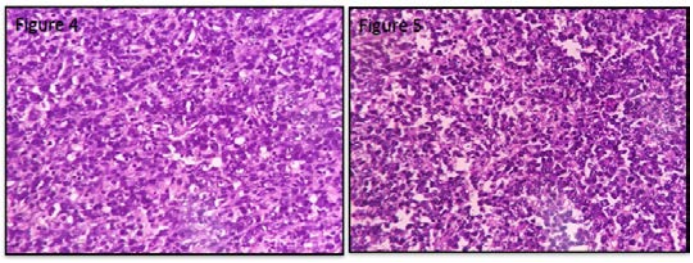
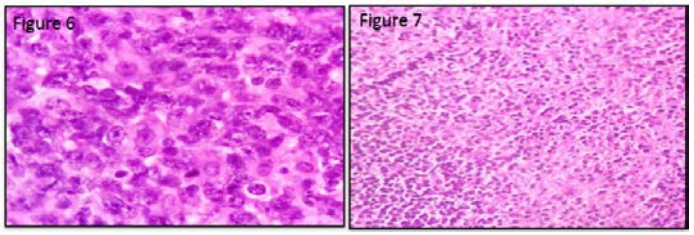


Figure 4, 5. Microscopic photograph (H & E, X 40)- left adrenal mass A,B – microscopic photograph showing sheets of medium to large sized neoplastic lymphoid cells with vesicular nuclei showing pleomorphism mitotic figure and areas of necrosis.



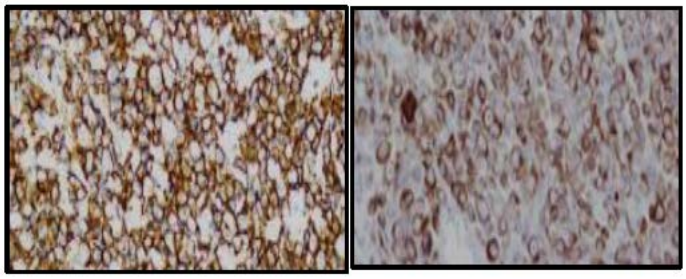


Figure 8. IHC markers (CD 20X 600), (CD79a X600) tumour cells showing CD 20 and 79a positivity