Original Research Paper

Volume - 10 | Issue - 8 | August - 2020 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Surgery

PRIMARY TESTICULAR LYMPHOMA: A RARE NEOPLASM

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ABSTRACT Primary Diffuse Large B Cell Lymphoma (DLBCL), although uncommon in the general population, is the most common type of malignant testicular neoplasm seen in males of older age group Its differential diagnosis are classical seminoma, viral and granulomatous orchitis. The typical clinical sign is a unilateral, painless testicular mass of variable size. Hereby, we present a case of primary DLBCL testis which was initially diagnosed as classical seminoma left testis involving left spermatic cord. The histological slides alongwith immunohistochemistry were further reviewed and categorized as primary DLBCL of testis. Its standard treatment has been with orchiectomy alongwith combination chemotherapy; but very few cases have a prolonged disease-free survival. Differential diagnosis includes seminoma and viral and granulomatous orchitis. Therefore diagnosis of this entity and appropriate management is very crucial.

KEYWORDS : Testis, Lymphoma, Primary, DLBCL

INTRODUCTION:

Primary testicular lymphoma constitutes about 1–9% of testicular tumors, most common in males more than 50 years of life. [1,2] DLBCL is the most common subtype of lymphoma (80–90%). It presents as a unilateral painless testicular mass of variable size. Treatment is orchidectomy followed by combination chemotherapy. DLBCL of testis does not have a favourable prognosis with tendency of local relapse and metastasis. Differential diagnosis includes seminoma, viral and granulomatous orchitis. Hence, a detailed histological examination and immunohistochemical correlation is essential to achieve at a diagnosis.

CASE REPORT

A 54 year old male presented with complaints of left testicular pain associated with fever since four months. He was a known hypertensive and a known diabetic for the last 4-5 years and on regular medications. There was no other associated history of Coronary artery disease/ Chronic obstructive pulmonary disease/ Kochs or any other drug allergy. On general examination, he was conscious, oriented, pulse rate was 80/minute, BP 130/90 mm Hg. He was afebrile and there was no pallor, cyanosis, edema, lymphadenopathy or icterus .Systemic examination was within normal limits. Local examination showed a hard and enlarged left testis, alongwith thickened spermatic cord. Hematological investigations revealed .Hemoglobin 13.2gm/dl, Total leucocyte count was 8300/mm^3 Polymorphs 68% Lymphocytes18% Eosinophils 8% Monocytes 06%. Urine routine microscopy was in normal limits. Total serum bilirubin was 0.6 mg/dl, direct being 0.2mg/dl, SGOT was 8 IU/L, Alkaline phosphatase was 348 U/L. Total protein was 6.6gm/dl, serum albumin 3.2 gm/dl and SGPT was 17 IU/L. Ultrasonogram of scrotum revealed that left testis was enlarged in size, 53x 37x 35mm with multiple hypoechoeic rounded areas, largest measuring 27x29x28mm Left epidydimis also showed hypoechoeic structures. Left scrotal sac showed presence of free fluid. Both testes showed tiny calcification. Right testes was normal in size and echotexture. A provisional diagnosis of infective etiology in left testis and epidydimis, ? Tubercular was made. However a repeat ultrasound was done which indicated malignancy of left testis. Human chorionic gonadotrophins level was <1.10 mIU/mL, alpha fetoprotein 2.87 IU /ml. Contrast enhanced computed tomography Scan abdomen revealed presence of reteroperitoneal lymphadenopathy with caseations within, left epidydimoorchitis and funiculitis, mild hydrocoel, possibility of Koch's etiology.

Patient underwent high inguinal orchidectomy and finally histopathology confirmed it initially as case of classical seminoma left testis involving left spermatic cord (stage pT3b, IPI 2/5) Patient was discharged on fifth post operative day on oral antibiotics and analgesics. Patient was referred to oncology department for further management and chemotherapy. The slides were further reviewed and on immunohistochemistry CD 20 was positive in large lymphoid cells, a final diagnosis of Non-Hodgkins Lymphoma, Diffuse large B-cell lymphoma left testis was made. The patient received three cycles of CHOP regime and fourth cycle of Intrathecal Methotrexate chemotherapy. The patient withstood the procedure and was discharged in a hemodynamically stable condition. Post chemotherapy, PET /CT imaging of whole body showed Non-FDG avid residual nodal mass, in left paraaortical region measuring 2.1x1.91cm in size which was previously measuring 5.1x5.8cm suggesting complete metabolic resolution of nodal masses. There was marked regression in size of previously seen nodal mass. No new lesion was seen, suggestive of good response to therapy.

DISCUSSION

In the recent years an increasing trend is seen in the incidences of primary testicular lymphoma and diffuse large B-cell lymphoma (DLBCL) is the most common testicular lymphoma accounting to more than 70% of the reported cases. Other types of testicular lymphomas comprise of follicular lymphoma, plasmacytoma, and lymphoblastic and Burkitt-like lymphomas [3,4] Recent trends have shown in the studies that the mean age of primary testicular lymphoma has shifted to younger age group, owing to increase in the number of HIV positive patients. [5,6]

Primary testicular DLBCL mimics the germinal cell tumours such as classical seminoma, spermatic seminoma and embryonal germ cell tumours.[7] DLBCL is considered to be an aggressive tumour which usually presents as a unilateral, painless, rapidly growing testicular mass alongwith involvement of extranodal sites [8C,9,10] Around 25-41% of patient present with constitutional symptoms like fever, anorexia, night sweats, and weight loss [11,12]

The scrotum and regional retroperitoneal lymph nodes are mostly involved alongwith local involvement of epididymis, spermatic cord as seen in the present case where left epidydimal involvement as well as reteroperitoneal lymph node involvement was seen.

On ultrasound, this tumour usually appears as focal hypoechoic mass without a definable capsule or diffuse enlargement when it is compared with the contralateral normal testis where hyperechoic areas are seen.[11] In the present case also such hypoechoeic rounded areas were obvious in both left testis as well as epidydimis.

Those patients who have limited disease, orchidectomy is helpful as it removes a sanctuary site, as the blood-testis barrier makes the testicular neoplasm inaccessible to chemotherapy.

Cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) regimen has been the main line of treatment for last several decades and recently anti-CD20 monoclonal antibody rituximab has been added to the CHOP regime which has immensely increased the survival.[13]

Testicular malignancy , especially in patients presenting at a younger age may mimic germ cell tumours Hence after histopathological evaluation an immunohistochemical confirmation is always recommended as lymphoma is an important differential diagnosis. The panel of immunohistochemical stains must include keratin, CD45, CD20, and OCT4. Therefore, it is very important to identify and

diagnose this entity correctly as there is a high risk of extranodal relapse even in cases with localized disease, moreover there are differences in line of treatment, management, and prognosis.

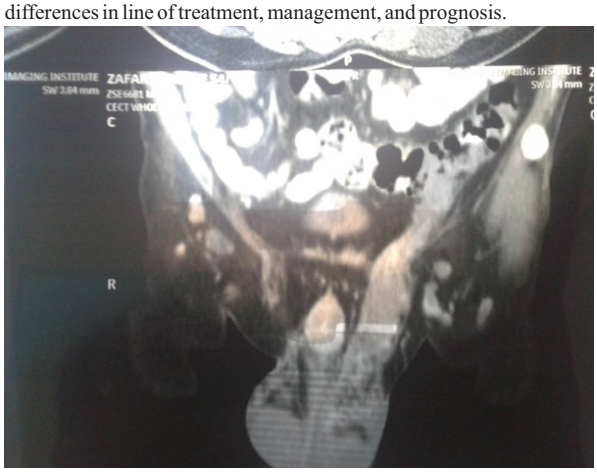

FIG 1: CECT FILM SHOWING TESTICULAR TUMOUR WITH MULTIPLE HYPOECHOEIC ROUNDED AREAS

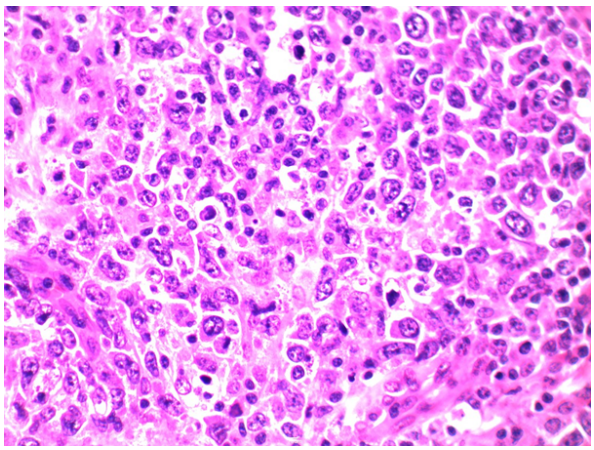

FIG 2: MICROPHOTOGRAPH SHOWING DIFFUSE GROWTH PATTERN WITH LARGE CELLS RESEMBLING IMMUNOBLASTS WITH CENTRAL NUCLEOLI AND CENTROBLASTS AND ATYPICAL MITOSIS(H &E,40X)

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