



Non-Hodgkins lymphoma (DLBCL type) of extranodal site: A rare case report.

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ABSTRACT

Non-Hodgkins lymphoma (NHL) are heterogenous group of neoplastic disorder of lymphoid tissue. It can occur as primary lymph node disease or at extranodal sites. Extra-nodal sites of involvement can be gastrointestinal tract, lung, thyroid, salivary glands, skin and female genital tract. We report a rare case of extra-nodal Non-Hodgkins lymphoma in a 40 year old female who presented with pain and swelling in abdomen. She was admitted due to a finding of a large retroperitoneal tumor in the lower abdomen and pelvis with a heterogenous appearing uterus which on histopathology and immunohistochemistry turned out to be diffuse large B cell lymphoma. She then received medical management, as is appropriate for this diagnosis. Chemotherapy was initiated in a very timely manner and she showed significant improvement within weeks.

Keywords: Non-Hodgkins lymphoma, Extra-nodal, Immunochemistry, Chemotherapy.

INTRODUCTION

Lymphomas are a diverse group of malignancies of the immune system. Non-Hodgkins lymphomas are a group of neoplasms that arises from the cells of the lymphoreticular system. Extranodal site involvement accounts for 25-30% of Non-Hodgkins lymphoma cases. Gastrointestinal tract is the most frequent site followed by the head and neck region constituting 11-33% of the cases. [1] The types of lymphomas encountered vary widely from one extra-nodal site to another.

Diffuse large B-cell lymphoma is the most common extranodal lymphoma. It is the most common lymphoma encountered in the central nervous system, including the eyes, paranasal sinuses, waldeyer ring, bone, heart, adrenals, and testis. It is the most common type of lymphoma encountered in the gastrointestinal tract and the female genital tract and it also occurs in the salivary glands, thyroid, mediastinum, orbit, oral cavity and other sites. [2]

CASE REPORT

A 45 year old female came with complaints of pain and swelling over abdomen since 3 months. It was associated with nausea and vomiting. CT scan showed a large retroperitoneal tumor mass with

invasion of uterus. Serum glutamic pyruvic transaminase (SGPT) was increased. A clinical diagnosis of leiomyosarcoma was kept. Patient underwent surgery and tumor was sent for histopathological examination along with hysterectomy specimen.



Fig. 1: Gross specimen of uterus with cervix with unilateral mass.



Fig. 2: Gross specimen of retroperitoneal mass



Fig. 3: Gross specimen of omentum with nodular deposits.

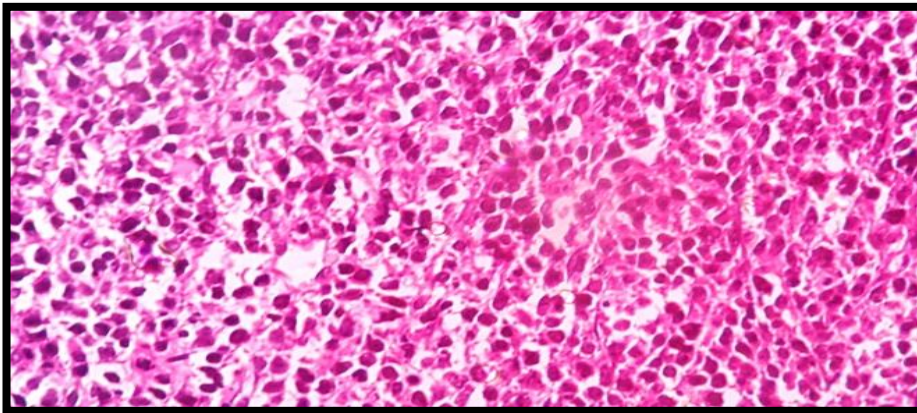


Figure 4: Section from tumor mass showing sheets of small to medium size cells with scanty cytoplasm and large oval hyperchromatic nucleus with coarsely clumped chromatin and prominent nucleoli.

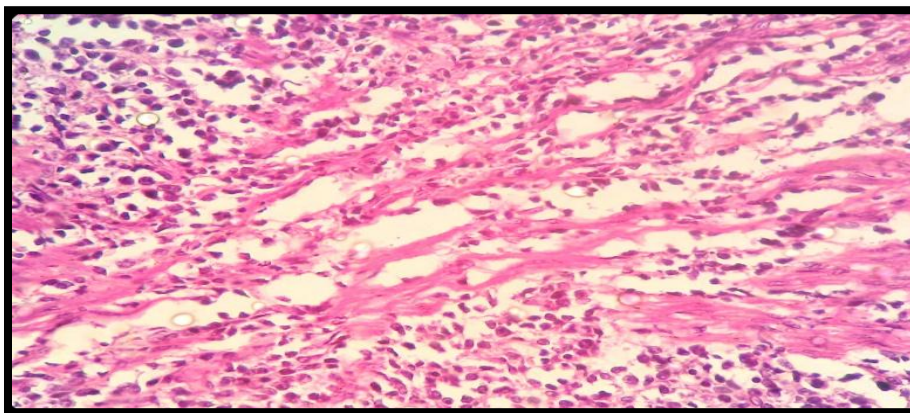


Figure 5: Section from myometrium with infiltration of uterine muscle with sheets of tumor cells.

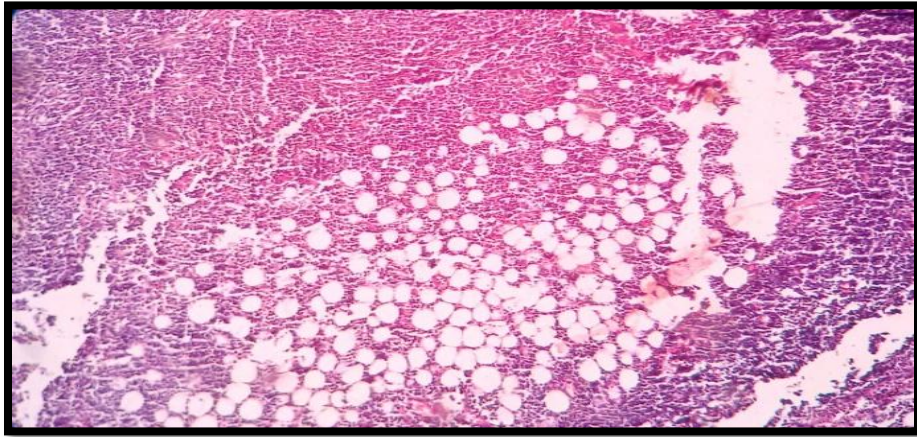


Figure 6: Section from omentum with adipose tissue showing infiltration by tumor cells.

On histopathological examination, differential diagnosis of Round cell sarcoma (probably soft

tissue sarcoma), lymphoma and endometrial stromal sarcoma was kept.

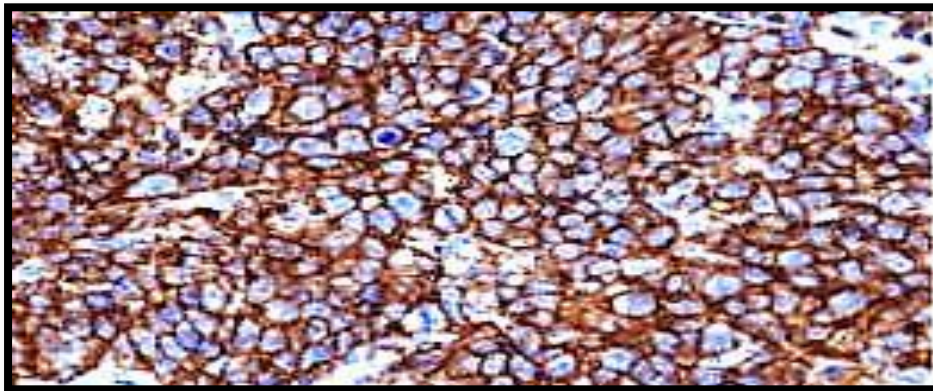


Figure 7: Leukocyte common antigen (LCA) positivity

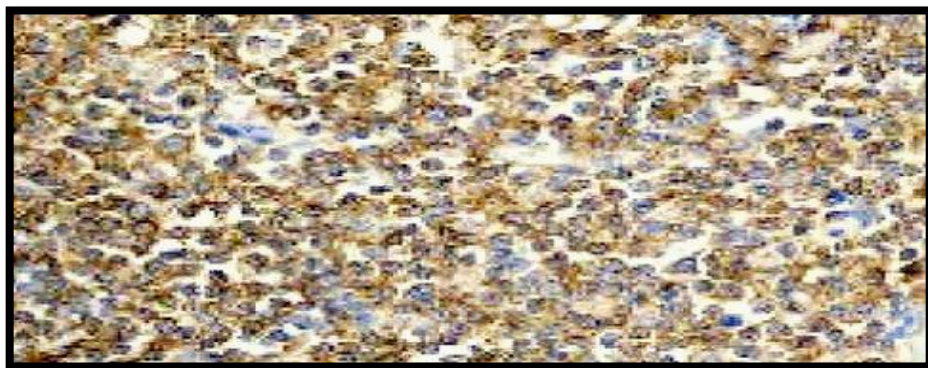


Figure 8: CD20 positivity.

Immunohistochemistry revealed tumor cells positive for LCA and CD 20 and immunonegative for cytokeratin, EMA, CD3,CD5,CD10 and Cyclin D1 leading to final diagnosis of Non-Hodgkins lymphoma of diffuse large B cell phenotype.

DISCUSSION

Extranodal lymphoma occurs in approximately 40% of all patients with lymphoma and has been described in virtually all organs and tissue.

Extranodal disease is more common with non-Hodgkin's lymphoma (NHL), and diffuse large B-cell lymphoma (DLBCL) is the most common histological NHL subtype in adults, accounting for ~25% of all NHL cases. Thus, it is known that gastrointestinal DLBCL is the most frequent form of extranodal lymphoma. However, DLBCL primarily arising in the retroperitoneal region has been rarely reported. The initial presentation of NHL varies depending on the subtype and involved area, with symptoms including enlarged palpable lymphadenopathy, B-symptoms (fever, weight loss, night sweats), and symptoms secondary to compression of adjacent structures. [3]

In our case, Patient presented with abdominal pain and mass. The neighbouring retroperitoneal organs are often involved by the tumor. Chen L et al in a study of 32 cases stated 37.5% of retroperitoneal lymphomas of diffuse large B cell type. [4] Some infectious agents have been associated more or less strictly with the development of a NHL as i.e., the Epstein-Barr virus, the human herpesvirus 8, and the human T-cell lymphotropic virus type [5].

NHL can present in the female genital tract, as an extranodal tumor or as ovarian, uterine, cervical, vaginal, vulvar or even fallopian tube as the primary site. The prevalence of these female genital tract lymphomas tend to be much lower, with rates seen to be between 0.2% and 1.1% in separate studies. Lymphoma of the retroperitoneum and pelvis can therefore be due to a nodal primary, a female genital tract primary, or from other abdominal organs. Extranodal lymphoma has been identified in all of the gynecologic organs and presents a significant diagnostic challenge due to the fact that it typically presents with symptoms

typical of traditional gynecologic primary pathology. These tumors are rare. Patients with NHL of the vagina, cervix, or uterus tended to present with pelvic pain, pelvic mass, and/or vaginal bleeding. Patients with cervical lymphoma may be diagnosed on cervical cytology, and this diagnosis can also be made at the time of examination for abnormal bleeding. Cervical lymphoma typically occurs in premenopausal women. Uterine lymphomas are more common in postmenopausal women and usually present with bleeding and the usual histology is large B cell lymphoma. Vaginal lymphoma typically presents with an obstructing mass, bleeding, or dyspareunia. These patients tend to do well with radiation or chemotherapy, however with advanced stage disease only one third were able to achieve long term remission in a previous study. Patients with vulvar lymphoma tend to be the oldest and have the worst outcomes despite aggressive multimodal therapies. Early indolent tumors are generally treated with radiation therapy alone and are typically nodular or follicular in origin. For patients with diffuse B cell lymphoma such as in the case presented, the current first line therapy remains to be Rituximab in combination with Cyclophosphamide, Doxorubicin, Vincristine and Prednisone (R-CHOP). [6]

CONCLUSION

Non-Hodgkins lymphoma of diffuse large B cell type presenting as retroperitoneal mass and involving uterus and cervix is very rare and should always be considered in such cases as early diagnosis and management is essential for better prognosis.

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