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Primary Non-Hodgkin's Lymphoma of Ovary

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Abstract: *The involvement of the ovary in lymphomatous processes is rare. Such an involvement can occur in 2 ways, either primary or secondary, which usually presents with abdominal or pelvic complaints. The distinction is of considerable importance because primary extra-nodal lymphomas run a less aggressive course. We present a case of primary diffuse large B-Cell lymphoma of ovary. The diagnosis was confirmed on histopathological examination and immunohistochemical analysis.*

Keywords: *Ovary, Non-hodgkins lymphoma.*

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I. Introduction

Involvement of ovary by malignant lymphoma is well known as a late manifestation of disseminated nodal disease, almost always of non hodgkins type but primary NHL are unusual [1]. Primary ovarian NHL accounts for 0.5 % of all NHL and 1.5 % of all ovarian neoplasms [2]. The most common histologic types involved in primary ovarian NHL are Burkitt lymphoma and diffuse large B-cell lymphoma [3]. Only a few cases have been reported yet and is worth reporting because with appropriate therapy, prognosis is favorable in these patients.

Case report

A twelve year old female patient attended the outpatient department (OPD) complaining of pain and heaviness in lower abdomen and fatigue for four months. On clinical examination, an ill-defined firm to hard large mass was palpable in the lower abdomen. There was no obvious presence of free fluid. The patient was anemic, normotensive and had generalized poor health. The liver and spleen were not palpable. Pelvic and digital rectal examination were normal.

Investigation

Hematological profile of the patient was normal except for hemoglobin level of 6.5 gram %. RBC morphology was microcytic hypochromic, ESR was increased being 75 mm in the first hour. Computed tomography scan showed heterogeneously enhancing soft tissue mass of 8.4 x 6.4 cm in the right adnexal region extending upto right lumbar region. There were no enlarged lymph nodes. The serum tumor markers were positive with CA-125 being 380 U/ml and serum LDH being 1460. Bone marrow aspiration was done which came out to be normal.

Management

An exploratory laparotomy was performed which revealed right sided ovarian tumor of 8.0 x 5.8 cm. The tumor was greyish white in color homogenous and firm in consistency. There was no residual normal ovarian tissue. Right sided oophorectomy was done. The abdomen was closed after peritoneal toileting. The entire operated specimen was sent for histopathological examination.

Pathological findings Grossly, the right sided ovarian tumor measured 8.4 x 6.0 x 3.0 cm with an intact capsule. The tumor was greyish white in color, homogenous and firm in consistency. The cut surface was fleshy.

Microscopically, dissections from right sided ovarian tumor showed a monotonous population of tumor cells in sheets. The cells were uniform and round with scanty cytoplasm, conspicuous nucleoli and clumped chromatin (Fig 1). There was increased mitotic activity. A thin rim of compressed ovarian tissue was also present. Immunohistochemically, the cells were positive for LCA and negative for cytokeratin(Fig 2), which confirmed the lymphoid origin of tumor.. The tumor cells were positive for B-lineage marker CD-20 (Fig3). The tumor cells were negative for T lineage marker, CD-3 and TdT. The Tumor cells showed a high Ki-67 index of 90% (Fig 4). On the basis of microscopic findings supported by IHC; a diagnosis of NHL of the ovary, Diffuse Large B-cell type (high grade) was made.

Figure 1-hpe of primary ovarian lymphoma(400x;with scanner view in inset)

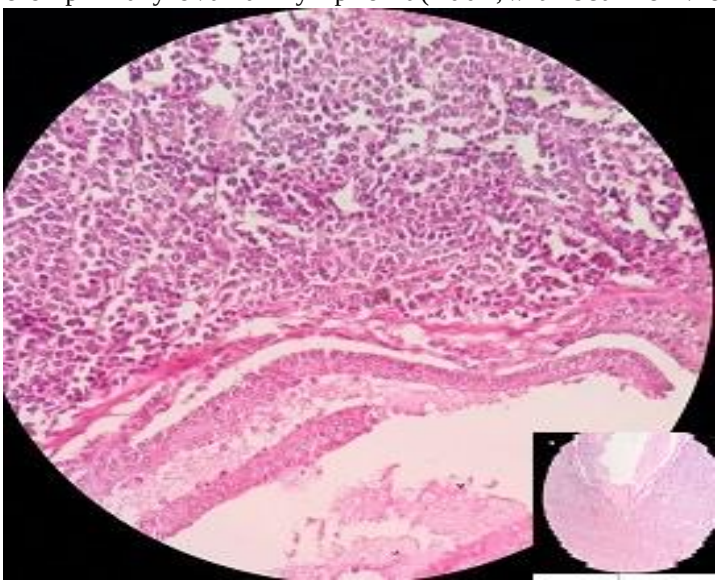


Figure 2-IHC showing negative for cytokeratin

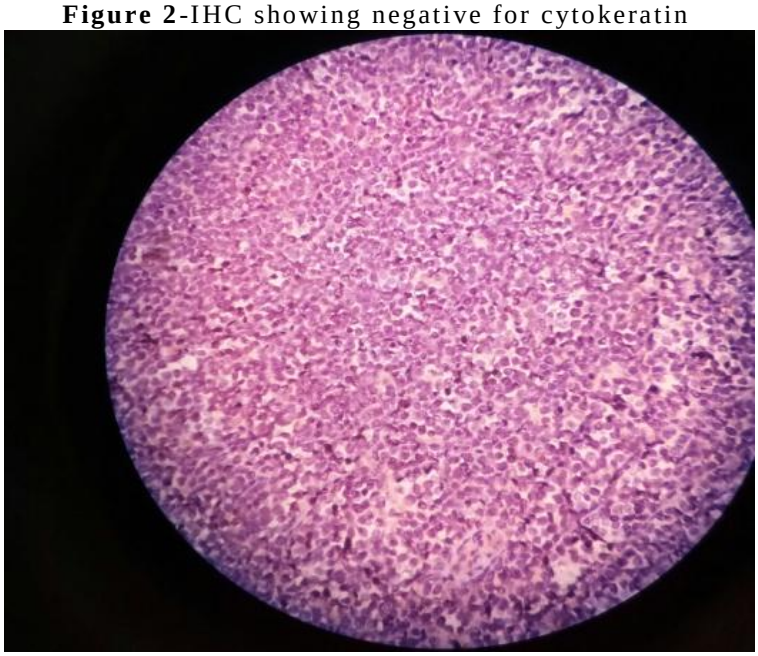


Figure 3-IHC showing positivity for CD20

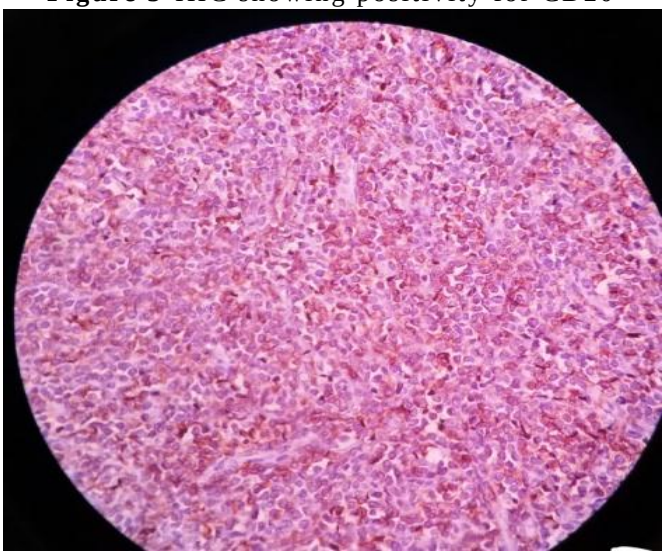
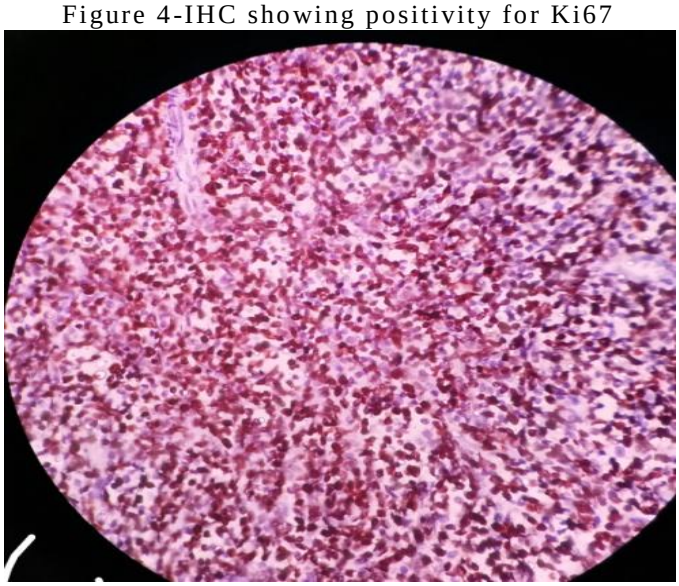


Figure 4-IHC showing positivity for Ki67



II. Discussion

NHL rarely involves the gynecologic tract. However, when involved ovary is one of the more common sites to be involved [4]. There has been debate as to whether lymphoma can arise de novo in the ovary; lymphoid aggregates do exist in normal ovarian tissue, which could give rise to such lesions [5]. The most common histologic types involved in primary ovarian NHL are Burkitt and Diffuse Large B-cell Lymphoma. Lymphomas of the ovary may occur at any age, but mostly women in their 40 s are affected [6]. The most common presenting signs or symptoms of malignant lymphomas involving the ovaries are abdominal or pelvic pain or mass [7]. Some cases present with ascites and elevated serum CA-125 [8]. Our patient presented with a pelvic mass and elevated serum CA-125 at the time of surgery. The presence of positive staining for LCA in the histological specimen distinguishes malignant lymphoma from non-lymphoid neoplasm [9]. These cells are negative for cytokeratin.

Lymphomas may simulate undifferentiated carcinoma or granulosa cell tumor on low magnification. Numerous IHC markers are available to identify these tumors as lymphoma [3]. In our case on microscopic evaluation, a differential diagnosis of NHL and granulosa cell tumor was made. Definite pathologic diagnosis was performed after immunohistochemistry. The presence of positive staining for LCA distinguishes malignant

lymphoma from non-lymphoid neoplasms. In our case, the tumor cells were positive for LCA and CD-20. Negativity of CD3 also confirms that it is of B-Cell origin.

The question of whether some ovarian lymphomas can be considered truly primary in the ovary and not merely a localized initial manifestation of a generalized disease cannot be answered yet. But Fox et al. [10] have suggested three criteria for the diagnosis of primary ovarian lymphoma.

At the time of diagnosis, the lymphoma is clinically confined to the ovary and a full investigation fails to reveal evidence of lymphoma elsewhere. A lymphoma can still, however, be considered as primary if spread has occurred to immediately adjacent lymph nodes or if there has been direct spread to infiltrate immediately adjacent structures.

The peripheral blood and bone marrow should not contain abnormal cells.

If further lymphomatous lesions occur at sites remote from the ovary, then at least several months should have elapsed between the appearance of the ovarian and extra-ovarian lesions. Our case was obviously primary ovarian malignant lymphoma, not a part of systemic disease. Because our case fulfilled all the criteria, we operated the patient for right ovarian solid mass. After the operation, whole body screening was found to be normal.

III. Conclusion

Primary ovarian lymphoma is a rare entity. It is frequently diagnosed as ovarian carcinoma which causes a significant delay in diagnosis and management. Best treatment option seems to be chemotherapy. Physicians should be aware of this rare entity to avoid radical surgical resection which seems to be unnecessary. More studies are required to better define and treat this rare entity.

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