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CASE REPORT

Rare ovarian neoplasm: Primary precursor B-cell Lymphoblastic Lymphoma involving bilateral ovaries

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Abstract

The involvement of the ovary in lymphomatous processes is rare. However in the female genital tract, the ovary is a frequent site to be involved by the hematological malignancies. Involvement of the ovary by malignant lymphoma can be primary or secondary and is discovered incidentally during a workup for abdominal or pelvic complaints. Most commonly occurring ovarian lymphoma is diffuse large B cell type, whereas the Precursor B Cell Lymphoblastic Lymphomas are extremely rare and previously only 5 cases have been reported. Here, we report a case of clinically suspected malignant ovarian tumor involving bilateral ovaries, which was diagnosed as a primary precursor B-LBL after surgery in a young female. This case highlights the need for careful evaluation of radiologic and morphologic features along with an extensive immunohistochemical panel to arrive at the correct final diagnosis to guide the chemotherapy.

Keywords: Non Hodgkin Lymphoma; Primary Ovarian Precursor B-Cell Lymphoblastic Lymphoma; Primary ovarian lymphoma; Primary ovarian tumors

Highlights

26 year old asymptomatic female with right adnexal mass underwent laparotomy and ovarian mass excision. She was diagnosed to have primary ovarian lymphoma by histopathology and radiology, concluded the final diagnosis of Primary Precursor B-cell Lymphoblastic Lymphoma by immunohistochemistry. This is the sixth reported case of Primary Precursor Lymphoblastic Lymphoma.

Introduction

Lymphomas presenting as ovarian tumors are uncommon and may occur de novo or secondary as a part of systemic disease. Primary ovarian lymphoma accounts for 0.5% of all Non-Hodgkin lymphomas and 1.5% of all ovarian neoplasms. Of which diffuse large B cell is commoner, whereas the precursor lymphoblastic lymphomas are extremely rare.[(1)]

Case History

A 26-year-old, primiparous woman, asymptomatic, underwent routine ultrasonography for pain abdomen of 3days duration and was found to have a pelvic mass. Her routine hematology and serum chemistry tests were all within normal limits. Further, contrast enhanced computed tomography (CECT) abdomen and pelvis scan confirmed the ultrasonography findings of large hypodense enhancing lobulated mass lesion measuring about 16.8x14.7x10.1cm in the right adnexa extending into the lower abdomen, whereas the left ovary appeared normal. Based on clinical and radiological data, the possibility of germ cell tumor was considered.

The patient underwent exploratory laparotomy and intraoperative frozen section of right ovary was suggestive of malignant ovarian neoplasm with a differential diagnosis of small cell carcinoma and Non Hodgkin Lymphoma (NHL). Therefore she underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and bilateral pelvic and para-aortic lymph node dissection. Peritoneal washings was also sent for fluid cytology. Her postoperative period was uneventful.

Grossly, the right ovary showed a circumscribed lobulated, solid grayish white mass, totally measuring 17x13.5x6.5cm and weighing about 850gm with attached fallopian tube. The cut surface was firm, fleshy and variegated, with complete replacement of the normal ovarian parenchyma. The left ovary was mildly enlarged. Microscopic examination of both ovaries showed starry sky pattern and complete replacement of ovarian architecture by round to oval intermediate-sized tumor cells arranged in a diffuse and cord-like arrangement. The tumor cells had high nuclear cytoplasmic ratio, convoluted nuclei with membrane folding, delicate chromatin, small nucleoli, scant cytoplasm and frequent mitoses (Figure 1). Right fallopian tube and omentum were also positive for tumor cell infiltration (Figure 2). Pelvic and paraaortic lymph nodes, left fallopian tube and peritoneal fluid were uninvolved. Histochemically tumor cells were negative for periodic-acid Schiff stain (PAS). Immunohistochemical analysis showed positive reaction for CD10, Bcl-2, CD99, terminal deoxynucleotidyl transferase, CD79a and Ki-67>95%. In contrast, tumor cells were negative for CD45 (leukocyte common antigen), CD20 (Figure 3), CD3, CD5, synaptophysin, chromogranin & pan-cytokeratin (Table 1). Thus, a final diagnosis of precursor B-LBL was considered.

Peripheral blood smear, bone marrow aspirate and biopsy examination revealed no evidence of leukemia or myeloproliferative disorder and ruled out systemic NHL. An elaborate clinical and radiological work-up did not reveal any lymphadenopathy, hepatomegaly or splenomegaly. Final diagnosis of primary bilateral ovarian precursor B-LBL was confirmed. Patient was treated with 6 cycles of CHOP regimen chemotherapy and follow PET-CT scan showed 2 years disease free survival after diagnosis.

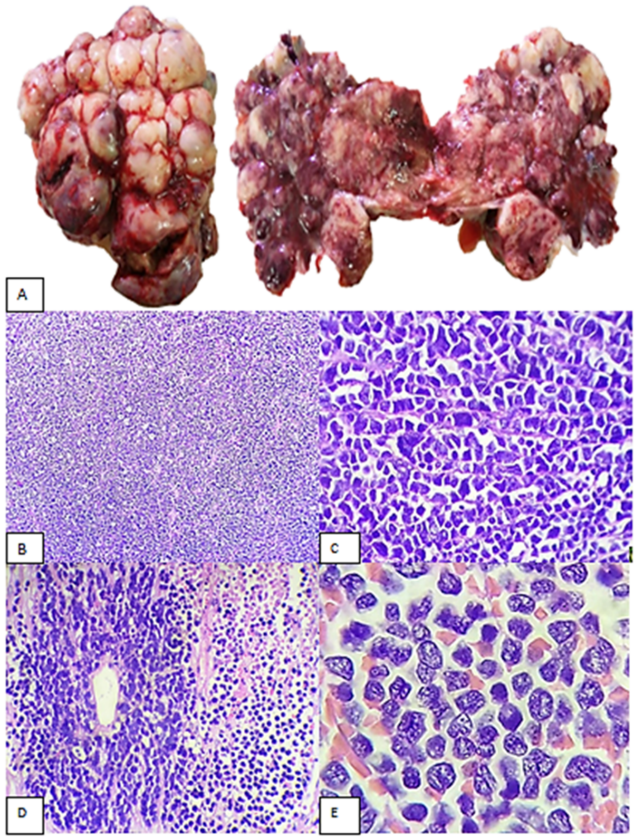

Fig 1. A. grossly, tumor is solid, firm, lobulated, variegated with grayish-white replacing the whole of right ovary. B. Ovarian parenchyma replaced by monomorphic tumorcells arranged in a diffuse pattern,starry sky appearance. C. Cord-like arrangement(single file pattern). D. Survival pattern- viable tumor cells around blood vessel. E. Individual tumor cells have high nuclearcytoplasmic ratio, convoluted nuclei with membrane folding, delicate chromatin, small nucleoli, scant cytoplasm and frequent mitoses

Discussion

NHL uncommonly involves the gynecologic tract. However, when involved by NHL, the ovary is one of the more common anatomic sites. [2] Lymphomas of the ovary, whether primary or secondary, have a varied presentation and are discovered incidentally during the work-up for pelvic and abdominal complaints. [7] The histogenesis of primary ovarian NHL is perplexing, as the normal ovaries are devoid of any lymphoid tissue. Therefore, benign lymphoid aggregates seen in the ovarian hilum and the chronic inflammatory cells found in the ovaries in response to pelvic inflammatory diseases have been suggested as the possible origins of primary ovarian lymphoma (POL). In fact, Monterroso et al. detected well-defined aggregates of lymphocytes in normal ovaries. [5]

In 1976, Fox et al. [8] proposed the following criteria to be fulfilled for the diagnosis of POL (with is very rare):

Table 1. Details of previously reported cases of primary ovarian lymphoblastic lymphoma in literature

Study	Age (yrs)	Symptom	Side of tumor	Diagnosis	IHC (positive)	IHC (negative)
VANG et al. [2]	29	Pelvic pain	R	T-LBL	CD3,CD10,CD99	CD20, CD21, BCL-6
PIURA et al. [3]	16	Lower pain abdomen	B/L	B-LBL	B LINEAGE	
IYENGAR et al. [4]	46	Abdominal distension	B/L	B-LBL	Tdt, CD43, CD45, CD10	CD5, CD8, CD20, CD79a, CK, VIM, CG, SYN, CD56
SAKURAI et al. [5]	19	Dysuria	R	B-LBL	Tdt, HLA-DR, CD10, CD20, CD79a, BCL-2, BCL-6	CK, VIM, CD45, CD3, CD5, CD15, CD30, CYCLIN D1
YADA et al. [1]	28	Incidental	B/L	B-LBL	Tdt, HLA-DR, CD34, CD99, CD20, CD79a, PAX5, CD43, MPO, bcl2, VIM	LCA, CD3.CD5, CD7, CD10, BCL-6, mum1, CD15, ALK, CD30, CD117, PLAP, CD68, CG, SYN, CK, EMA, INH, CALRET
JOEL et al. [6]	18	Lower pain abdomen	L	B-LBL	LCA, CD20, C79a, CD10, TdT, Ki67>90%	LMO, EMA, CD3, CD68
PRESENT CASE	26	Incidental (pain abdomen)	B/L	B-LBL	CD10, BCL2, Ki-67>95%, CD99, CD79a, Tdt+++	CD45, CD20, CD3, CD5, SYN, CG, PAN CK

B/L, bilateral; B-LBL, precursor B lymphoblastic lymphoma; CD, cluster of differentiation; CK, pancytokeratin; CALRET, calretinin; CG, chromogranin; HLA, human leukocyte antigen; EMA, epithelial membrane antigen; INH, inhibin; LCA, leukocyte common antigen; MPO, myeloperoxidase; mum1, multiple myeloma oncogene 1; PAX5, paired box 5; PLAP, placental alkaline phosphatase; R, right; RT, radiotherapy; SYN, synaptophysin; Tdt, terminal deoxynucleotidyl transferase; T-LBL, precursor T lymphoblastic lymphoma; VIM, vimentin.

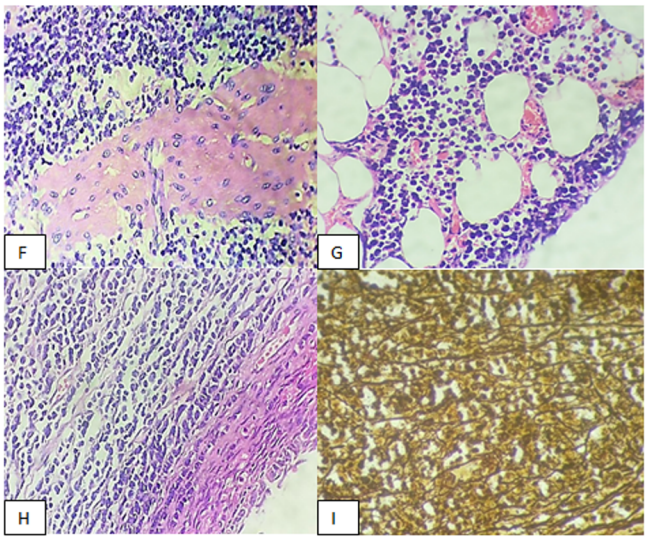

Fig 2. F. Right fallopian tube and G. Omentum infiltrated by similar tumor cells. H. Left ovary with corpus luteal cyst and similar tumor cells.

(i) The lymphoma is clinically confined to the ovary and a complete investigation fails to reveal evidence of lymphoma elsewhere at the time of diagnosis. However, an ovarian lymphoma can still be considered as primary if it has spread to immediately adjacent lymph nodes or if it has directly spread to infiltrate immediately adjacent structures.

(ii) The peripheral blood and bone marrow should be free of tumor cells.

(iii) In case of lymphomatous lesions occurring at sites remote from the ovary, atleast several months should have been elapsed between the appearance of the ovarian and extraovarian lesions.

The present case has fulfilled all the above criteria.

Examination of various surface and nuclear markers of lymphoma is necessary when malignant lymphoma is suspected by hematoxylin and eosin staining of sections. The microscopic features of ovarian lymphoma are similar to those seen in the extra-ovarian sites, except that there is a great tendency for the tumor cells to grow in cords and nests, appearing as to cling to the reticulin, forming pseudoacini which create a diagnostic dilemma. The most common types of lymphomas encountered in the ovary are Diffuse large B-cell lymphoma(DLBCL), Burkitt lymphoma(BL) and follicular lymphomas(FL); however precursor B-LBL is rare. [7] Out

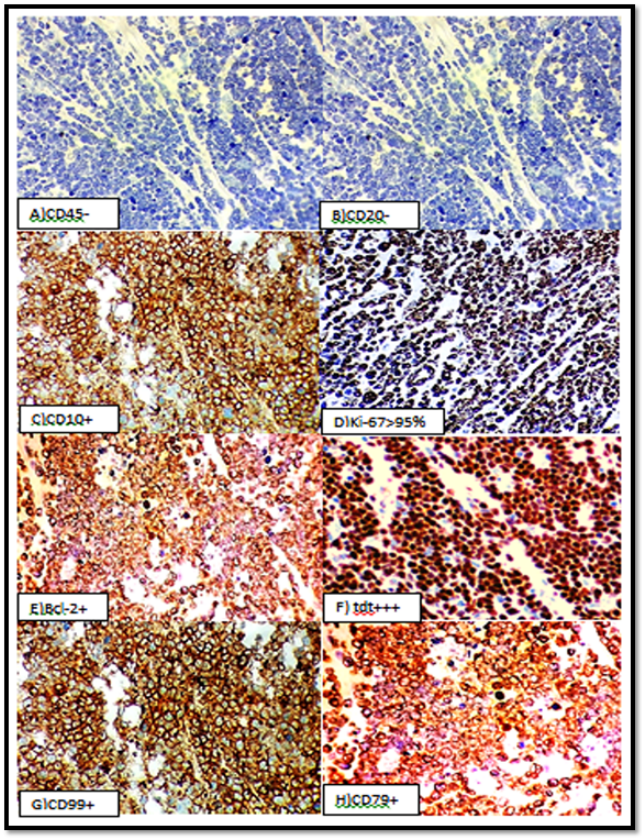

Fig 3. A. CD45-negative. B. CD20-negative. C. CD10-positive. D. Ki-67>95%. E. BCL-2-positive. F. tdt-strong positive. G. CD99-positive. H. CD79a-positive

of the five cases of primary ovarian precursor lymphoblastic lymphoma described in the published in English literature (Table 1), one was of T cell lineage.[2] Piura et al. reported a case of precursor B-LBL in a patient aged 16 years.[3] Iyengar et al. described a case with focal bony involvement of the lymphoma which was observed after surgery and chemotherapy.[4] The third case has been described by Sakurai et al. in a 19-year-old woman presenting with ovarian tumours and para-aortic lymphadenopathy.[5] Yadav et al described a case of primary precursor B cell LBL with aberrant expression of myeloid marker.[1] It should be noted that diffuse infiltration of the adjacent fallopian tube is much more common in lymphomas than in most of the tumors in the differential diagnosis.[9]

We ruled out histologically similar neoplasms mimicking ovarian NHL including granulosa cell tumor, dysgerminoma, small cell carcinoma with hypercalcaemia, granulocytic sarcoma, primitive neuroectodermal tumor and rhabdomyosarcoma with the help of immunohistochemistry. Since the tdt was strongly positive, BL and DLBCL are also ruled out.

To conclude, primary ovarian lymphoma of the precursor B-LBL type is extremely rare, usually asymptomatic when detected and poses a diagnostic dilemma. Therefore, a careful evaluation of radiologic and morphologic features, such as cord-like pattern of tumor cells, along with an extensive immunohistochemical panel, is required for diagnosis. Patients with localized disease and B cell phenotype have a good prognosis.

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