

Primary Non-Hodgkin Lymphoma Of The Ovary: A Rare Occurrence

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Abstract:

(1) Non-Hodgkin lymphoma (NHL) involving the ovary is uncommon, constituting only about 1.5% of ovarian tumours. It can arise in two forms:

Primary ovarian NHL (PONHL): This is a rare type (0.5% of NHL) where the lymphoma originates in the ovary itself.

Secondary ovarian NHL: This is more frequent, occurring in 7% to 26% of disseminated lymphomas, where the ovary is affected by a systemic disease.

The most common subtype of NHL in the ovary is diffuse large B-cell lymphoma. Patients may present with pelvic pain, a mass, or other non-specific symptoms. Diagnosis can be challenging as it can mimic epithelial ovarian cancer. Treatment typically involves chemotherapy, with radiotherapy as a possible addition.

Keywords: Non-Hodgkin lymphoma, ovary, primary, secondary, diffuse large B-cell lymphoma

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I. Introduction:

Non-Hodgkin lymphoma (NHL) is a heterogeneous group of lymphoid malignancies affecting the lymphatic system. While NHL can involve various organs outside the lymph nodes, its presence in the ovary is uncommon accounting for 1.5% of ovarian malignancies and 0.5% of overall non-Hodgkin lymphoma. This case report presents a patient diagnosed with primary non-Hodgkin lymphoma of the ovary.

We will discuss the clinical presentation, diagnostic workup, treatment approach, and potential challenges associated with this rare ovarian malignancy. This case adds to the existing knowledge base on non-Hodgkin lymphoma of the ovary and highlights the importance of considering this diagnosis in patients presenting with suspicious ovarian masses.

II. Case Report

A 41-year-old, para2 woman presented with abdominal pain for about 2 months. No history of any mass per abdomen or fever, chills or weight loss. Her past medical history was not significant and she denied any family history of malignancies.

Physical examination revealed a mass arising from pelvis corresponding to gravity uterus of 20 weeks, on tender, freely mobile horizontally. Laboratory investigations showed mild anaemia with Hb of 9.4g% and no leucocytosis or thrombocytopenia. Elevated LDH levels at 1060U/ml. Elevated CA125-257U/ml. Other tumour markers including CEA, AFP, Beta-HCG were normal.

Pre operative CT scan showed a mass in pelvis arising from left annexation reaching till umbilicus measuring 8*11.6*13(APxTRxSI), left ovary not seen separately. Right adnexa showed a round lesion with fat attenuation and a focus of calcification likely dermoid cyst. Multiple homogeneously enhancing left paraaortic lymph node are noted, largest measuring 12mm in SAD.

Diagnosis and Treatment:

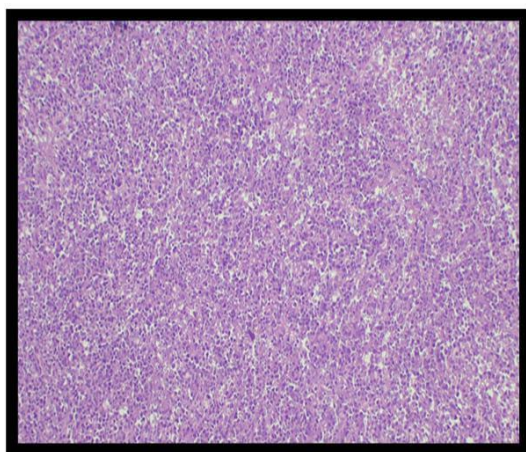
Given the patient's presentation and imaging findings, a differential diagnosis of ovarian malignancy was considered. Patient underwent laparotomy, intra-operatively a solid cystic mass 13*12*13cm noted to arise from left ovary. And right cyst measuring 5*6*5cm containing haemorrhagic fluid. No enlarged lymph nodes. Uterus and fallopian tubes were normal. Undersurface of diaphragm, paracolic gutter and pouch of Douglas were clear. Since Frozen section facility is not available in our institution, specimen was sent for histopathological examination.

Histopathological examination revealed non-Hodgkin lymphoma of left ovary with medium sized lymphoid cells with prominent nucleoli. No viable ovarian parenchyma noted in section with necrosis, right ovary showing dermoid cyst.

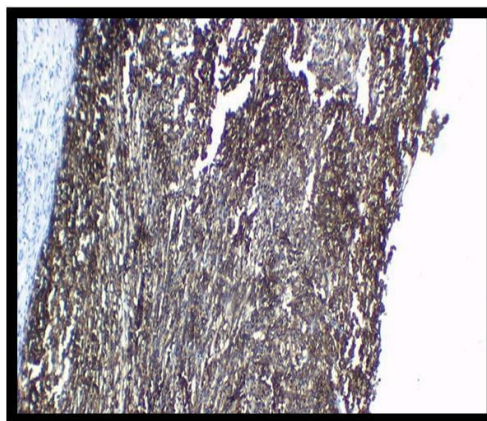
Gross specimen of left ovary showed a bosselated, irregular, shows presence of solid areas entirely. Cut section showing presence of yellowish solid areas with areas of haemorrhage band cystic areas.

Bone marrow biopsy showed no malignant cells. Immunohistochemistry shows cells weakly positive for CD45, BCL2, CD20(diffuse strong positive). Ki67 proliferation index is 25-30% in focal areas. Negative for WT1, INHIBIN, CD3, CD10, CD5, CD24, CYCLIN D1, TDT, CMYC, BCL6.

Diagnosis of non-Hodgkin lymphoma of left ovary was made.



H&E



CD20 (L26)

III. Discussion:

Non-Hodgkin lymphoma (NHL) involving the ovary is an uncommon presentation of a blood cancer. This discussion will delve into the key aspects of ovarian NHL, highlighting its challenges and management strategies.

Rarity and Forms:

NHL constitutes a mere 1.5% of all ovarian malignancies.

It manifests in two forms:

Primary Ovarian NHL (PONHL): Extremely rare (0.5% of NHL cases), arising from lymphoid tissue within the ovary.

Secondary Ovarian NHL: More frequent (7-26% of disseminated NHL), where the ovary is affected by a systemic lymphoma.

(2) Diagnostic challenges:

Clinical Presentation: Symptoms like pelvic pain, mass, or fatigue can mimic epithelial ovarian cancer, making early diagnosis difficult.

B Symptoms: Fever, night sweats, and weight loss are less common in PONHL compared to other cancers, potentially delaying suspicion of NHL.

Imaging Studies: Ultrasound and CT scans may not be definitive for diagnosis.

(3) Here are the established criteria by Fox et al. to diagnose PONHL:

Tumour confined to the ovary (including unilateral/bilateral ovaries) or regional lymph nodes/structures at the time of diagnosis.

Bone marrow and peripheral blood smear free of any abnormal lymphoma cells.

No evidence of extraovarian disease at diagnosis. If lymphoma appears later, it could suggest a misdiagnosis of an earlier, more subtle disseminated disease.

In contrast, secondary ovarian NHL will not meet these criteria. It will likely show evidence of disseminated disease elsewhere in the body.

Additional Considerations:

Frequency: PONHL is much rarer than secondary involvement.

(4) Prognosis: Generally, PONHL has a better prognosis compared to secondary NHL due to its earlier stage at diagnosis.

The rarity of ovarian NHL necessitates a high index of suspicion, particularly in patients with atypical presentations.

(5) A multidisciplinary team approach involving gynaecologists, oncologists, and pathologists is crucial for optimal management.

Further research is needed to improve diagnostic accuracy and develop targeted therapies for ovarian NHL.

IV. Conclusion:

Non-Hodgkin lymphoma of the ovary presents diagnostic challenges due to its rarity and overlapping features with other malignancies. A high level of clinical suspicion, thorough workup with tissue biopsy, and multidisciplinary care are essential for effective management and improved patient outcomes.

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