

Primary ovarian lymphoma — a rare entity

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Abstract

Primary ovarian lymphoma is very rare. Ovarian involvement by non-Hodgkin's lymphoma is usually secondary, occurring as part of a systemic disease. We report a case of primary diffuse large B-cell lymphoma of the ovary manifesting as advanced ovarian cancer. The diagnosis was confirmed on histological and immunohistochemical analysis. The patient was treated with surgery followed by chemotherapy using rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone (R-CHOP) regimen.

Keywords: Lymphoma, Ovary.

Introduction

Genital tract involvement in non-Hodgkin's lymphoma (NHL) is uncommon. However, when involved, the ovary seems to be one of the more common anatomic sites. Ovarian involvement is usually secondary, occurring as part of a systemic disease.¹ Localised primary lymphoma of the ovary is unusual and rare, with few reported cases.²⁻⁶ With appropriate therapy, prognosis is favourable.^{1,7}

Case Report

A 64-year-old, para 4+1, postmenopausal woman was admitted with complaints of weight loss and heaviness in the lower abdomen. On examination, a firm and fixed abdominal mass could be palpated from the pelvis up to the umbilicus; ascites positive on percussion. Computed tomography (CT) scan revealed a large 23x18x16cms solid right ovarian mass with involvement of the adjacent bowel, ascites and metastatic peritoneal deposits. Serum CA-125 was elevated (413 IU/ml).

On laparotomy, a large, solid tumour arising from the right ovary could be seen that involved the rectum and sigmoid colon (Figure-1). The right ureter was dilated with the lower part surrounded by the tumour. The left ovary and the uterus were normal. Multiple, firm, metastatic nodules were present in the omentum and on the peritoneal surface. Removal of tumour with hysterectomy and left salpingo-oophorectomy was performed along with omentectomy. Lower part of sigmoid and upper part

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Figure-1: Gross appearance of the removed solid ovarian tumour with the involved sigmoid colon, rectum, omentum, uterus and the other ovary.

of rectum was removed en bloc followed by end-to-end anastomosis. The tumour measured 30x20x20cms at the time of resection.

Histopathology of the ovarian tumour was reported to be consistent with diffuse large B-cell lymphoma, non-germinal centre like subgroup (2008 WHO Classification) based on morphological and immunohistochemical features⁸ (Figure-2). Tumour cells were reported positive

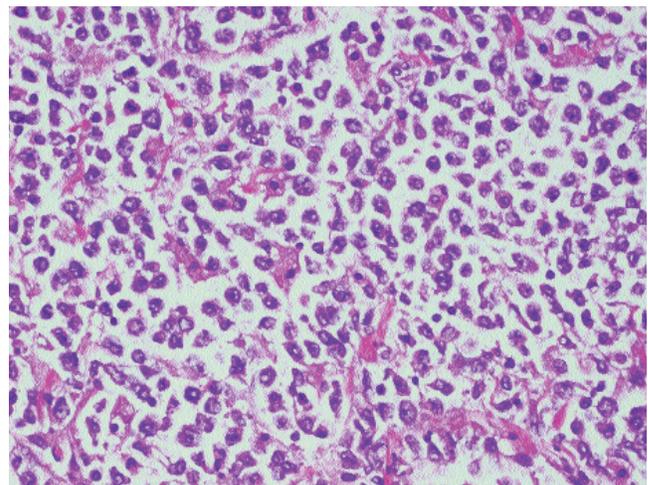


Figure-2: Histopathology showing diffuse large B-cell Lymphoma.

for CD79a, CD20 and BCL-6. Ki67 was positive, showing high proliferative index.

The patient had bulky extranodal disease involving the ovary stage I EBX (E - involvement of extranodal site; B - presence of B symptoms i.e. fever, weight loss more than 10% body weight; X - bulky disease more than 10cms in size) according to the Ann Arbor System.^{6,9} International prognostic index (IPI) reading was 2 (good risk). Post-operatively, the patient was given chemotherapy using the rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone (R-CHOP) regimen.

Discussion

Ovarian involvement by malignant lymphoma is well-known as a late manifestation of disseminated nodal disease. However, primary ovarian lymphoma as the initial manifestation is rare⁵ accounting for 0.5% of all NHLs and 1.5% of all ovarian neoplasms.⁷ Malignant lymphomas involving the ovaries are seen at autopsy with a frequency of 7-26%.⁸

Primary ovarian lymphomas present more often with symptoms attributable to an ovarian mass and are usually larger than secondary tumours.¹ The neoplasms may be microscopic or very large, and are usually unilateral. Some cases present with ascites and elevated serum CA-125.^{2,9} Our patient presented with large, unilateral ovarian mass with ascites and elevated serum CA-125.

The differential diagnosis of solid ovarian tumours includes rhabdomyosarcoma, extragonadal teratoma, neurogenic tumour, granulosa cell tumour and dysgerminoma. Definitive diagnosis can only be confirmed by histological examination of the tumour tissue.⁸

The histogenesis of primary ovarian NHL is perplexing. In normal ovaries, lymphocytes of B-cell and T-cell lineage are present within cortical granulomas and rare lymphocytes are dispersed throughout the ovarian stroma and within ovarian follicles and corpora lutea.¹⁰ Both B-cell and T-cell NHLs can arise in the ovary. Diffuse large B-cell lymphoma appears to be the most common type of primary ovarian NHL.²

The histology in our case was reported to be consistent with diffuse large B-cell lymphoma of the ovary. Tumour cells were reported positive for CD20 and BCL-6. Ki67 was positive, showing high proliferative index. A series of eight cases of primary ovarian NHL reported three of them to be diffuse large B-cell lymphoma positive for CD20 and BCL-6.²

The use of chemotherapy is based on the principle that ovarian lymphoma must be considered a localised manifestation of the systemic disease. The prognosis for such patients is much better than that of patients with obvious systemic disease. The protocol for chemotherapy used in diffuse large B-cell histology is the standard R-CHOP regimen.⁷ The prognosis for these patients, treated with appropriate chemotherapy, appears to be similar to that of patients with other nodal NHLs.⁷

Conclusion

Primary lymphoma of the ovary is a rare entity. It is considered a localised manifestation of the systemic disease. The treatment principles and prognosis are the same as that of other nodal NHLs. With appropriate management, the outcome is favourable.

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