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Primary ovarian non-Hodgkin's lymphoma

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Summary

Background: Primary ovarian non Hodgkin's lymphoma (PONHL) is a very rare disease. We present a case of PONHL and review the literature. *Case:* The patient, a 24-year-old nulliparous Greek woman, presented with the complaint of abdominal pain. She underwent left salpingo-oophorectomy, multiple biopsies from the right ovary, total omentectomy, pelvic and paraortic lymphadenectomy, appendectomy and curettage. The histopathology revealed diffuse large B-cell non-Hodgkin's lymphoma of the left ovary. She underwent postoperative chemotherapy. She remains well without evidence of disease, 15 months after initial surgery. *Conclusion:* The use of chemotherapy is based on the principle that PONHL must be considered a localized manifestation of systemic disease. Patients with PONHL have a similar outcome compared to patients with other NHL.

Key words: Ovarian non-Hodgkin's lymphoma; Treatment; Chemotherapy; Prognosis.

Introduction

Primary ovarian non Hodgkin's lymphoma (PONHL) is a very rare disease [1, 2]. It accounts for 0.5% of all non Hodgkin's lymphomas (NHL) and only 1.5% of all ovarian neoplasms [3].

The median age at diagnosis of PONHL is 42 years (range 22-69 years) [3]. We present a case of PONHL and review the literature.

Case Report

The patient, a 24-year-old nulliparous Greek woman, presented with the complaint of abdominal pain. Her past surgical history was unremarkable. Her family history revealed no evidence of cancer among the first-degree relatives.

On gynaecologic examination there was a palpable and mobile pelvic mass. There were no palpable inguinal lymph nodes and the rest of pelvic examination was normal.

Preoperative magnetic resonance imaging (MRI) of the abdomen and pelvis, and abdominal ultrasound (US) revealed an intraabdominal mass of 17.5 x 10.5 x 4.5 cm, without enlarged lymph nodes. Preoperative computer tomography (CT) of the chest, chest X-ray, colonoscopy and cystoscopy were normal. Preoperative CA-125 was elevated at 71.4 U/ml and lactate dehydrogenase (LDH) was extremely elevated at 1200 U/l.

On exploratory laparotomy, the left ovary was markedly distended, measuring 17.5 x 10.5 x 4.5 cm. Frozen section showed malignancy and the patient underwent left salpingo-oophorectomy, multiple biopsies from the right ovary, total omentectomy, pelvic and paraortic lymphadenectomy, appendectomy and curettage.

Histopathology revealed diffuse large B-cell non-Hodgkin's lymphoma of the left ovary. The right ovary was normal. The omentum, all removed lymph nodes and appendix were negative for malignancy. Most of the malignant cells were CD-20 and leucocyte common antigen (LCA) positive. The peritoneal washing smear was negative for malignant cells. A bone marrow biopsy showed no abnormalities.

The final diagnosis was primary ovarian non Hodgkin's lymphoma Stage IE according to the Ann Arbor staging system [4].

The patient underwent postoperative chemotherapy. She received eight courses of the standard CHOP regimen (cyclophosphamide 750 mg/m^2, doxorubicin 50 mg/m^2, vincristine 1.4 mg/m^2 and prednisone 50 mg/m^2) in combination with Rituximab (375 mg/m^2).

Follow-up 15 months after initial surgery with CT of the chest, abdomen and pelvis, abdominal US, chest X-ray, IVP, colonoscopy, urethrocystoscopy and bone marrow biopsy showed no evidence of recurrence.

Discussion

Involvement of the ovary in NHL is well recognized [5]. However PONHL is a very rare disease [1, 2]. It accounts for 0.5% of all NHL and only 1.5% of all ovarian neoplasms [3]. This is partly because there is no lymphoid tissue within the ovary. It has been suggested that the tumour originates from lymphocytes in the ovaries surrounding blood vessels at the hilum and is related to the corpus luteum [1, 6]. It is possible that these lymphoid aggregates could give "de novo" rise to lesions as lymphoma [7].

The most common presenting signs or symptoms of PONHL are abdominal or pelvic pain or mass [1, 3, 8]. Other less frequent complaints are urinary incontinence, irregular vaginal bleeding, nausea or vomiting, and swelling of the lower extremities [1, 3]. Fever, night sweats or weight loss (B symptoms) were noted in 10%-33% of the patients [1, 3].

Bilateral ovarian involvement by NHL has been reported in 36-71% of patients [1, 3, 5, 9, 10]. Both B- and T-cell NHL can arise in the ovary, but B-cell PONHL are much more frequent (80-95%) [8, 11]. Among them diffuse large B-cell lymphoma is the most common type between 35 and 45 years [11], whereas follicular lymphoma and small lymphocytic lymphoma are more often found in older women [10, 12]. In our case, histopathology revealed diffuse large B-cell PONHL.

Fox and Langley [13] proposed the following diagnostic criteria for primary ovarian lymphoma:

1. At the time of diagnosis, the lymphoma is clinically confined to the ovary, and a full investigation fails to

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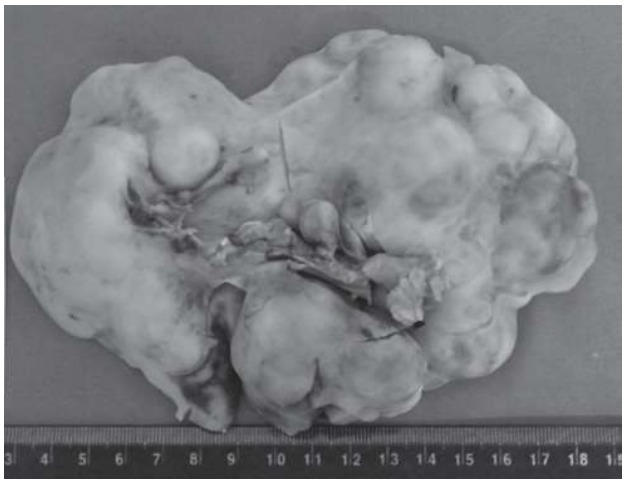
Fig. 1



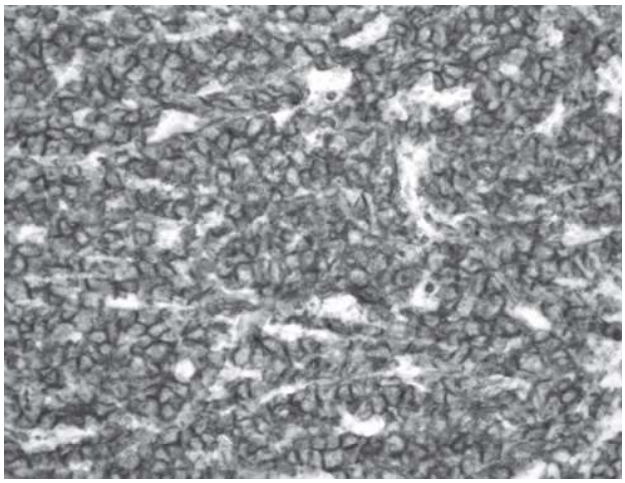


Fig. 2

Figure 1. — Macroscopic view of the ovarian tumour.
Figure 2. — Immunohistochemical positive result for CD-20 showed B cell lymphoma of the ovary (CD-20x400).

reveal evidence of lymphoma elsewhere. A lymphoma will still be considered primary if spread has occurred to immediately adjacent lymph nodes or if there has been direct infiltration of adjacent structures.

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

3. 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.

According to these diagnostic criteria, our case was PONHL.

Most patients with PONHL are treated with surgery followed by chemotherapy [1, 8, 10]. The use of chemotherapy is based on the principle that PONHL must be considered a localised manifestation of systemic disease [10]. A CHOP regimen is often administered, because CHOP is the most standard regimen for the treatment of NHL [3, 14, 15]. In our case, the patient underwent postoperative chemotherapy with the CHOP regimen in combination with rituximab.

The prognosis for patients with PONHL localised to one ovary is much better than that of patients with obvious systemic disease [1]. Patients with PONHL have a similar outcome compared to patients with other NHL [1, 3, 16].

In conclusion, the use of chemotherapy is based on the principle that PONHL must be considered a localised manifestation of systemic disease. Patients with PONHL have similar outcomes in comparison to patients with other NHL.

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