Non-Hodgkin’s lymphoma may involve the gynecologic tract, and the ovary is one of the commonest anatomic sites to be involved (1). However, ovarian involvement by non-Hodgkin’s lymphoma is usually – in approximately 90% of cases – secondary, occurring as manifestation of systemic disease. Localized, considered primary ovarian non-Hodgkin’s lymphoma (PONHL) is a very rare entity (2, 3). It accounts for 0.5% of all non-Hodgkin’s lymphomas (NHL) and only 1.5% of all ovarian neoplasms (4, 5).

The median age at diagnosis of PONHL is estimated at 42 years (range 6-69 years), something that leads to fertility preservation dilemmas in many cases of young patients. We present a rare case of bilateral primary ovarian non-Hodgkin’s lymphoma.

Case report: A 38-year old nulliparous woman, underwent exploratory laparotomy because of bilateral ovarian masses. Left salpingo-oophorectomy, partial omentectomy and excision of an ovarian mass of the right ovary was performed. Great effort in order to preserve healthy ovarian tissue of the right ovary as well as the right fallopian tube was given, due to fertility reasons. Final histology showed bilateral diffuse large B-cell primary ovarian non-Hodgkin’s lymphoma. Postoperatively, the patient underwent chemotherapy with the CHOP regimen in combination with rituximab. Five years after initial diagnosis, the patient remains well with normal menstrual cycle, without evidence of recurrence.

Discussion: Fertility preservation issues in some cases of rare gynecological malignancies could be managed via minimally invasive oncological approach.

**SUMMARY:** Bilateral primary ovarian non-Hodgkin’s lymphoma and fertility preservation: 5-year follow-up.

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Aim. Primary ovarian non-Hodgkin’s lymphoma is a very rare disease. Median age at diagnosis is estimated at 42 years, something that leads to fertility preservation issues in many cases. This was a case report study, presenting a rare case of bilateral primary ovarian non-Hodgkin’s lymphoma.

Case report. A 38-year old nulliparous woman, underwent exploratory laparotomy because of bilateral ovarian masses. Left salpingo-oophorectomy, partial omentectomy and excision of an ovarian mass of the right ovary was performed. Great effort in order to preserve healthy ovarian tissue of the right ovary as well as the right fallopian tube was given, due to fertility reasons. Final histology showed bilateral diffuse large B-cell primary ovarian non-Hodgkin’s lymphoma. Postoperatively, the patient underwent chemotherapy with the CHOP regimen in combination with rituximab. Five years after initial diagnosis, the patient remains well with normal menstrual cycle, without evidence of recurrence.

Discussion. Fertility preservation issues in some cases of rare gynecological malignancies could be managed via minimally invasive oncological approach.

**KEY WORDS:** Ovarian cancer - Ovarian lymphoma - Fertility preservation - Adnexal mass - Chemotherapy.
Discussion

Ovarian lymphoma is a very rare entity. The ovarian involvement by malignant lymphoma may be as primary extranodal disease and the secondary involvement may be as an initial manifestation of clinically occult nodal disease or as a late complication of disseminated nodal lymphoma (1, 6). Primary extranodal lymphoma is less aggressive as 5 years survival rate is estimated at 80%, whereas it is 33% in cases of secondary lymphoma (6, 7). This is why differential diagnosis between them is of great importance.

However, definition of PONHL remains debatable since long. Fox and Langley (8) 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 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.

The very low incidence of PONHL could be partly explained by the absence of 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 (2, 5, 9). It is possible that these lymphoid aggregates could give ‘de novo’ rise to lesions as lymphoma.

The most common signs or symptoms of PONHL are abdominal or pelvic pain or mass (1-4). Less frequent complaints are irregular vaginal bleeding, nausea or vomiting, urinary incontinence and swelling of the lower extremities (2, 3). B-symptoms (fever, night sweats or weight loss) were reported in 10-33% of the patients (2, 3). In our case the patient presented with the most common symptom; this of abdominal pain.

Both B- and T-cell NHL can arise in the ovary. However, diffuse large B-cell lymphoma is the most common subtype of NHL and the most common subtype of PONHL, as well (1, 6). The presence of positive staining for LCA in the histological specimen distinguishes malignant lymphoma from non-lymphoid neoplasm. Also, diffuse large B-cell lymphomas are positive for CD20 and negative of T-lineage marker CD3, such as in our case.

Bilaterality among PONHL is infrequent (1). However, in our case intraoperative findings and frozen section of the left adnexae and the mass of the right ovary were positive for bilateral malignant tumours. Great effort in order to preserve healthy ovarian tissue of the right ovary as well as the right fallopian tube was given, due to fertility reasons. Our patient was 38 years old, nulliparous and more conservative surgical approach was indicated in order to preserve ovarian function and fertility. Final histology of the specimen of the removed mass of the right ovary showed no invasion of the surgical margins. This minimally invasive management led our patient to a normal ovarian function with normal menstrual cycles after CHOP chemotherapy. Another similar case of bilateral ovarian Burkitt’s lymphoma in a 10-year old girl, in which fertility preservation surgical approach was followed, was described by Lee AC et al. at 2015 (10).

Most patients diagnosed with PONHL are treated with surgery followed by chemotherapy (1, 2). The use of chemotherapy is based on the principle that PONHL must be considered a localized manifestation of systemic disease. CHOP is the most standard regimen for NHL treatment (4).
Conclusions

Although rare, NHL should be included in the differential diagnosis of bilateral ovarian tumours. A minimally-invasive oncological approach in order to preserve ovarian function and fertility could be followed in cases of localized disease in young women and girls.

Conflicts of interest

All authors declare no conflicts of interest.

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