

Primary ovarian large b-cell lymphoma: a case reportSinaa Mohamed¹, Elkhanoussi Basma², Albouzidi Abderrahmane³¹Department of pathology, military hospital Moulay Ismail, Meknes, Morocco²Department of pathology, national institut of oncology, Rabat, Morocco³Department of pathology, military hospital Mohamed V, Rabat, Morocco***Corresponding author**

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Abstract: The involvement of the ovary by malignant lymphoma is a well-known late manifestation of disseminated nodal disease. Primary ovarian lymphoma is rare. We report a case of primary ovarian non-Hodgkin's lymphoma with unilateral involvement which was managed by surgery and chemotherapy. A 42-year-old woman was admitted with signs and symptoms suggestive of an ovarian cancer. Computed tomography revealed a tumor of the left ovary measuring 12 cm in diameter, without enlarged lymph nodes. The tumor was classified as a diffuse large B-cell lymphoma. The patient has been advised 6 cycles of CHOP regimen and is presently on treatment. She has now been without disease for 36 months after the surgery. According to previous reports the treatment principles and prognosis of primary ovarian large b-cell lymphoma is the same as that of other nodal lymphomas.

Keywords: large b-cell lymphoma, ovary, primary

INTRODUCTION

Involvement of the ovary by malignant lymphoma is well known as a late manifestation of disseminated nodal disease, almost always of the non-Hodgkin's type. But primary ovarian lymphomas are unusual [1]. Primary ovarian non-Hodgkin's lymphoma accounts for 0.5% of all non-Hodgkin's lymphomas and 1.5% of all ovarian neoplasms [2].

Fox and al. have suggested three criteria for the diagnosis of primary ovarian lymphoma: 1- tumor has confined to the ovary regional lymph nodes or adjunctive organs at the time of the diagnosis, 2- bone marrow and peripheral blood have not contained any abnormal cells, and 3- if extraovarian disease appear later, there must be a few months between the time of ovarian and extra-ovarian lesions [3]. The most common type of lymphoma involving the ovary is diffuse large B-cell lymphoma [4]. We present a case of primary ovarian large b-cell lymphoma, manifesting like ovarian cancer, which was managed by surgery and chemotherapy.

CASE REPORT

A 42-year-old woman (gravida 3, para3), was accused four months of pelvic pain, sometimes associated with leucorrhea, Otherwise no sign of compression associated. The pelvic exam and digital rectal examination were normal. The ganglionic areas were free from the disease. Computed tomography (CT) scan showed a unilateral heterogeneous tissular abdominopelvic tumor measuring 12 cm in the large diameter; there were no enlarged lymph nodes [Figure 1]. The serum tumor markers were positive, with CA-125 being 130 U/ml (normal < 25 U/ml). An

exploratory Laparotomy revealed a left ovarian tumor measuring 12x7 cm in diameter which was white, tissular, and without extracystic vegetations or ascetic fluid accumulation; there were no peritoneal implant, or adenomegaly. A unilateral adnexectomy with omentectomy were performed.

On gross examination, the left ovarian mass measured 12 × 7.0 × 4.0 cm with an intact capsule and was solid and grayish white. The omentum was covered by exudates and showed no nodules. Microscopic examination of ovarian mass revealed ovarian tissue was replaced by diffuse sheets of monotonous large cells [Figure 2]. The cells had a scant cytoplasm and cleaved nuclei-clumped chromatin. Neither follicular structures nor starry-sky pattern was noted. Immunohistochemically, the cells were positive for CD-20 [Figure 3]. This cells were negative for the T-cell marker : CD3 [Figure 4]. A diagnosis of diffuse primary large B-cell lymphoma involving the ovary was rendered.

Work-up for the lymphoma after the surgery included abdominal and thoracic magnetic resonance imaging, bone marrow biopsy, and a whole body positron emission tomography (PET) scan. The findings

from all these studies showed no other sites involved. The patient later received an adjuvant R-CHOP chemotherapy (rituximab, 375 mg/m² day; cyclophosphamide, 750 mg/m² day; doxorubicin, 50 mg/m² day; vincristine, 1.4 mg/m² day; prednisone, 50

mg/m² 2 day) 6 times intravenously and treatment progressed well. The patient is alive without disease 36 months after the operation without additional surgery.



Fig-1: Computed tomography scan show a unilateral, heterogeneous, tissular mass of the left ovary

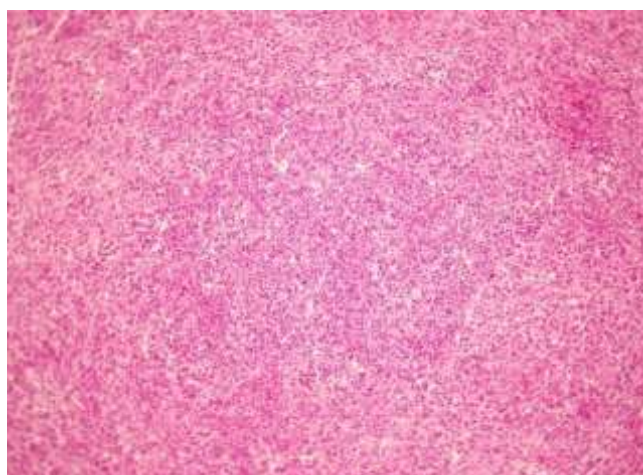


Fig-2: Hematein-eosin (HE) : Diffuse sheets of monotonous large cells, with a scant cytoplasm and cleaved nuclei-clumped chromatin (x100 magnification)

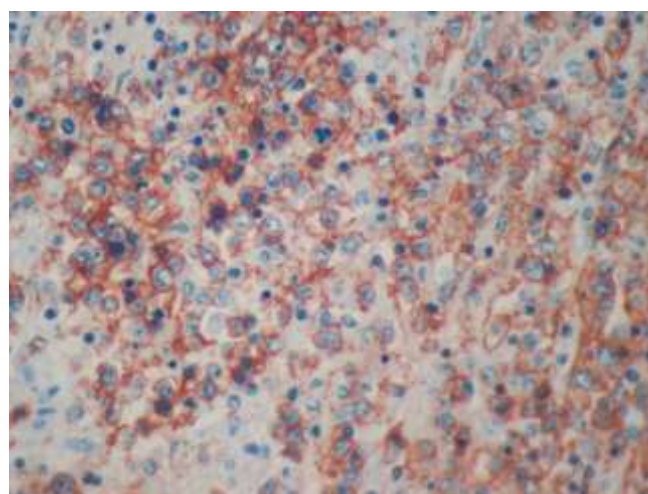


Fig-3: immunohistochemistry (IHC) studie showed strong positivity of the tumor cells for CD20. (x200)

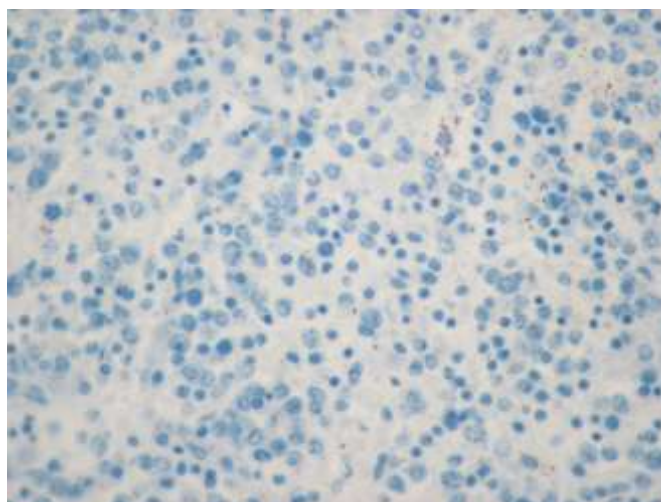


Fig-4: The tumor cells was negative for CD 3 (x200)

DISCUSSION

Non-Hodgkin lymphoma rarely involves the gynecologic tract. However, when involved, the ovary is one of the more common anatomic sites [4]. True primary ovarian lymphomas are even rarer. Primary ovarian NHL accounts for 0.5% of extranodal NHL and 1.5% of primary ovarian cancers [2]. The most common histologic types involved in primary ovarian NHL are Burkitt lymphoma and diffuse large B-cell lymphoma [5]. The differential diagnosis of solid ovarian tumors includes rhabdomyosarcoma, extragonadal teratoma, and granulosa cell tumor, and definitive diagnosis can be only confirmed by pathologic examination with immunochemistry study of the tumor tissue.

The presence of positive staining for lymphoid marker (CD20, CD 3) distinguishes malignant lymphoma from non lymphoid neoplasms. In our case, tumor cells were positive for CD20. It's were negative for CD 3.

The difference between primary and secondary lymphomas is important in terms of prognosis. The initial clinical manifestation of an occult nodal lymphoma as an ovarian mass is known as having a poor outcome with a survival rate ranging from 7% to 38% at 5 years. But primary ovarian lymphoma has better prognosis [6]. The histologic type is probably the most important prognostic factor, with B-cell tumors being associated with longer survival [7]. In our case, diffuse large B cell lymphoma of the ovary has a favorable outcome and the patient is alive without disease for 36 months.

Primary ovarian lymphomas are staged as other extranodal NHL (Ann Arbor staging system) [3]. Treatment of primary ovarian lymphomas is based on histology, type, and clinical staging. Patients with localized disease usually have better prognosis [8]. In

our patient, lymphoma was localized the unilateral ovary. Bone marrow biopsy, PET scan, and abdominal and thoracic MRI were clear. Ann Arbor stage was stage 1E. After the operation and clinic evaluation our patient was treated with 6 cycles of R-COP regimen of chemotherapy every 3 weeks. No recurrence could be detected during a followup of 36 months.

The question of whether some ovarian lymphomas can be considered truly primary in the ovary and not merely a localized initial manifestation of a generalized disease cannot be answered yet. But Fox *et al.* [9] have suggested three criteria for the diagnosis of primary ovarian lymphoma. If stringent criteria of Fox and al. are applied, primary ovarian lymphoma becomes vanishingly rare. Our case was obviously primary ovarian malignant lymphoma, not a part of systemic disease. Because our case fulfilled all the criteria, we operated the patient for left ovarian mass. There were no other pathologic findings about the pelvic organs. After the operation a whole body screening was clean.

Ferrozzi and *et al.* [10] reported eight patients with ovarian NHL (two primary lymphomas and six systemic NHLs) and assessed their most typical imaging patterns. Ovarian lymphomas were frequently bilateral and homogeneous, without ascites, and the tumors always exceeded 5 cm in diameter. Ultrasonography showed homogeneous, hypoechoic, and mildly vascularized tumors. Ultrasonographic features are nonspecific. In our case, ovarian mass was unilateral, heterogeneous. In the operation it was seen that the solid component was clear. So salpingo oophorectomy was performed. Patients with ovarian lymphomas are mostly diagnosed after surgery and are treated with chemotherapy [10] as in our case. Radiotherapy is optional.

CONCLUSION

Primary ovarian large b-cell lymphoma a very rare disease. It is diagnosed after the operation using immunohistochemistry and treated with chemotherapy. According to the previous reports in literature, the treatment principles and the prognosis are the same as that of other nodal lymphomas.

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