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Case Report

Primary Ovarian Burkitt's lymphoma Mimicking a Gynecologic Tumor

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Abstract

Ovarian non-Hodgkin's lymphoma is an extremely rare tumor. It accounts for 1.5% of ovarian cancers and 0.5% of non-Hodgkin's lymphoma (NHL). It is most often a secondary location of a disseminated lymphomatous disease. We report on a patient managed in the department for this condition.

Keywords: Ovarian lymphoma, pelvic masse, immunochemistry, chemotherapy.

INTRODUCTION

Burkitt's lymphoma (BL) was first described by Burkitt in 1958 [1]. It is a highly aggressive non-Hodgkin lym- phoma, often presenting in extranodal sites or as an acute leukemia [1]. Adult patients with sporadic BL often present with extranodal disease, and the abdomen is the most frequent site of involvement [2]. However, malignant lymphomas involving the female genital tract are rare, and <1% of patients initially present with ovarian enlargement [3]. Although ovarian lymphoma is a disease with poor prognosis, it is reported that successful results are possible with chemotherapy and surgery [3]. Nevertheless, an extranodal BL presenting with signs and symptoms mimicking a primary gynecologic tumor is extremely rare. In this article, we present a case of BL mimicking a gynecologic malignancy. In addition, the patient did not have the common "B" symptoms associated with lymphoma, such as fever, night sweats, and fatigue.

CASE REPORT

We report the case of a 40 years old patient, operated at the age of 13 for appendicular peritonitis, admitted for chronic pelvic pain without other associated signs, the abdominal examination finds a fixed pelvic mass of 10*08cm, of hard consistency, the gynaecological examination finds an irregular bulging mass at the level of the posterior vaginal cul de sac. The pelvic ultrasound objects the presence of bilateral latero-uterine polylobed tissular masses measuring 11*8,5cm on the right and 08*08cm on the left without effusion, completed by a pelvic MRI revealing a voluminous median pelvic mass lateralized on the left of 12*8cm which infiltrates the uterine body and the right ovary, also invades the left ureter responsible for a ureterohydronephrosis laminating the homolateral renal cortex and encompasses the vessels without obstructing them. This mass appeared to continue with a poorly bounded, irregularly contoured, left- sided sub- and retroperitoneal tissue infiltrate measuring approximately 96 x 150 x 133 mm in diameter. CA125 was 46 IU.

A CT scan revealed locally advanced left ovarian tumor process, 10*17cm, responsible for left ureterohydronephrosis, with left external iliac and inguinal adenopathies. Thickening of the peritoneal sheets with infiltration of the perilesional fat, probably related to the carcinosis. Repressed right ovary with hydrosalpinx. Pulmonary abnormalities suggestive of infectious disease such as pulmonary tuberculosis and mediastinal lymph nodes were also present.

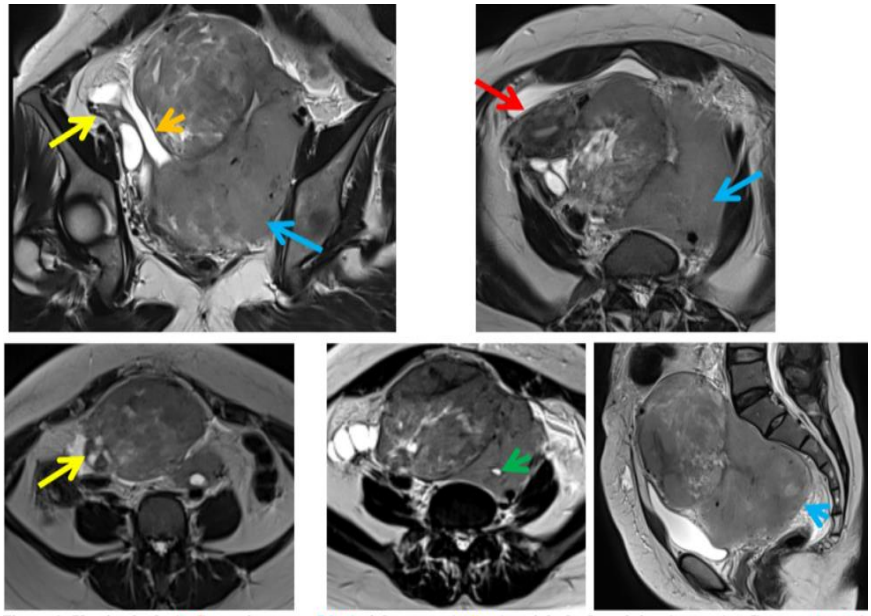

Figure 1: Uterine body (red) ovarian mass (blue) right ovary (yellow) + right hydrosalpinx (orange) + G ureter (green)

The patient was operated, we found an enormous mass, suprauterine, highly vascularised, with anarchic vascularisation, without buds or vegetation on the surface, probably at the expense of the non-visualised left ovary pushing back the uterus (of normal size) anteriorly and laterally to the right, this mass seems to have a deep invasion in the sub-peritoneum, non-visualised right ovary probably invaded by the tumor with right hydrosalpinx of 07cm.

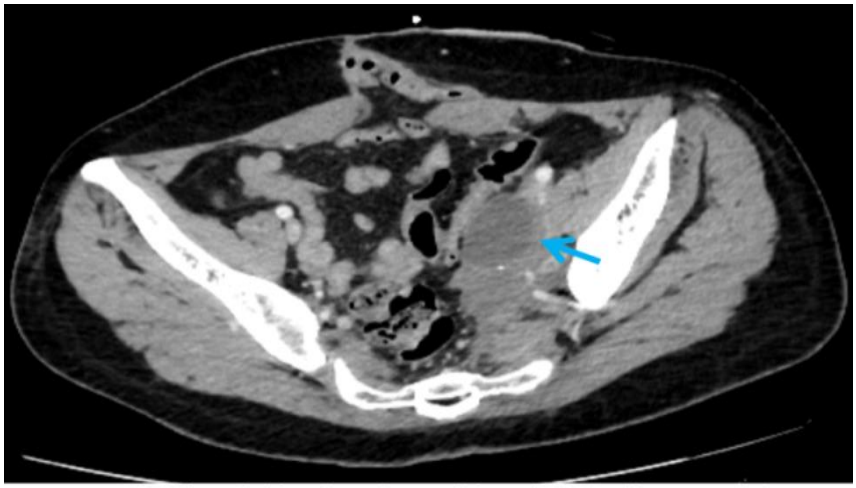

Figure 2: After chemotherapy: left ovarian mass (clear regression)

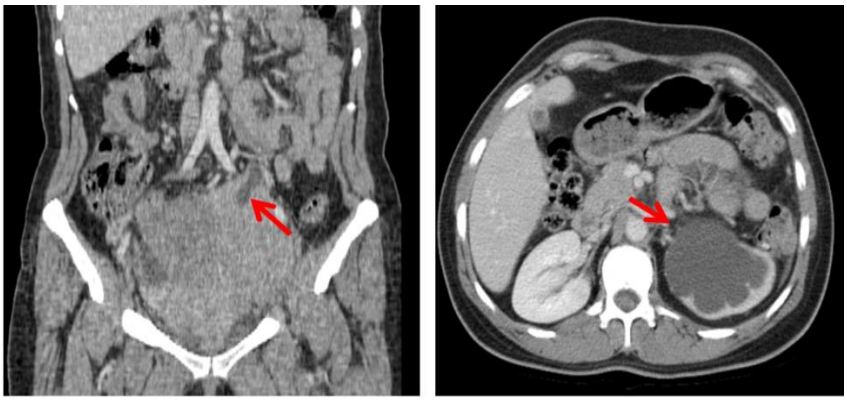

Figure 3: Invasion of the left ureter with significant upstream uretero-pyelic ectasi

The decision was to resect the tumor, which was referred for extemporaneous examination revealing tumor proliferation in favor of a granulosa tumor, with accidental removal of the left ureter, which was engulfed by the friable tumor, the urologist resected a part of the left ureter and percutaneous ureterostomy has been established.

The immunohistochemical study was compatible with a diffuse large cell B lymphoma of ovarian location: Ac anti CD 20: +, Ac anti CD 3:-, Ac anti PLAP ET INHINE –, pax 5 +, Cd10-, Ac anti Ki67: 60 %, Ac anti CD 5: -, Ac anti CD Bcl 6: -, Cycline d1 -, Ac anti CDMUM1: -.

Our patient received a COP then 8 RCHOP treatments with a very good therapeutic response estimated at 93% after 4 cures.

On the follow-up CT scan after 4 cures, there was a clear regression in size of the large left latero-uterine pelvic tissue mass of ovarian origin, currently measuring 8x4.6 cm in diameter versus 15x13 cm in diameter, with complete regression of the invasion of the right pelvic ureter and of the dilatation of the upstream excretory cavities, Hypotrophic left kidney, normal parenchymography, without dilatation of its excretory cavities with drainage probe along the pyelon and right ureter directed towards the anterior abdominal wall in relation to the ureterostomy setup.

On the PETSCAN there was not a suspicious looking hypermetabolic abnormality, especially in projection of the left latero-uterine mass and adnexal collection.

DISCUSSION

Involvement of the ovary in malignant lymphoma is well known as a late manifestation of disseminated nodal disease. However, primary ovarian lymphoma with secondary ovarian involvement as an initial manifestation of lymphoma is unusual.

The ovary is the most frequent site of non-Hodgkin's lymphoma in the female genital tract, and Burkitt lymphoma has been reported to account for approximately 19% of adnexal lymphomas [4]. Fox and Langley proposed the following criteria for the diagnosis of a primary ovarian lymphoma:

- The first criterion is the clinical confinement of the lymphoma to the ovary and a lack of evidence of lymphoma elsewhere on complete investigation at the time of diagnosis. A lymphoma can still, however, be considered as primary if spread has occurred to immediately adjacent lymph nodes or if there has been direct spread to infiltrate immediately adjacent structures.
- The second criterion is that the peripheral blood and bone marrow should not contain any abnormal cells.
- Third, 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 [5].

There are two forms of Burkitt lymphoma: endemic and sporadic. The endemic form shows head–neck lesions. However, these lesions were not noted in this case, which was therefore considered to be of the sporadic form.

Malignant lymphoma is revealed by an ovarian mass in less than 1% of all non-Hodgkin lymphoma. Primary ovarian lymphoma (POL) is still much rarer and represents 0.5% of NHL and 1.5% of all ovarian neoplasms [6]. Diffuse lymphoma and a B-cell phenotype are the most common histologic type and phenotype, respectively. Lymphoma may indeed involve the ovary as a primary neoplasm, as an initial clinical manifestation of an occult nodal disease, or that of a widely disseminated systemic lymphoma. The distinction between primary and secondary lymphomas is usually made postoperatively, when the diagnosis of ovarian lymphoma is established and when its extension is evaluated [6].

In the case of an ovarian mass, few signs suggest the diagnosis of ovarian lymphoma. Most patients are young, with a median age of 42–47 years. Fever, emaciation, or night sweats have to be investigated during the patient's interview. Clinical examination will seek a palpable adenopathy, or a liver or spleen infiltration. Our patient was 40 years old, she has a chronic pelvic pain with pelvic mass of 10*08cm.

Biologically, A decrease of peripheral red blood cells or circulating lymphomatous cells can be found, as well as an inflammatory syndrome, a deficient immune response, usually with hypogammaglobulinemia, sometimes associated with a monoclonal peak. A tumoral lysis syndrome can also be found [6].

Ferrozzi *et al.*, reported eight patients with ovarian NHL (two primary lymphomas and six systemic NHL) 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. In all cases, computed tomography (CT) revealed clear-cut hypodense lesions, with mild contrast enhancement. MRI revealed homogeneous masses, with low signal on T1-weighted images and slightly hyperintense on T2-weight images, and gadolinium T1-weighted images showed a mild

enhancement. Moreover, the concurrent finding in the literature of adenopathy or hepatosplenomegaly can confirm the diagnosis, and a compression of the urinary tract is often reported [7].

In the case we report, the pelvic ultrasound objects the presence of bilateral latero-uterine polylobed tissular masses measuring 11*8,5cm on the right and 08*08cm on the left without effusion, completed by a pelvic MRI revealing a voluminous median pelvic mass lateralized on the left of 12*8cm which infiltrates the uterine body and the right ovary, also invades the left ureter responsible for a ureterohydronephrosis laminating the homolateral renal cortex and encompasses the vessels without obstructing them. This mass appeared to continue with a poorly bounded, irregularly contoured, left-sided sub- and retroperitoneal tissue infiltrate measuring approximately 96 x 100 x 133 mm in diameter, there was not ascites.

Macroscopically, the tumor is usually large, smooth, soft, and friable. Its color is often reported to be pale gray as the case of our patient.

The diagnosis of LMNH of the ovary can only be made on histology, thus all lymphoma phenotypes can be observed in the ovary. Diffuse large-cell B-cell lymphoma, as in our case, is the most frequent phenotype. Indeed, the histological appearance of ovarian LMNH sometimes leads to the discussion of many diagnoses in view of the indifferent appearance of proliferation, including granular cell tumour, dysgerminoma, hypercalcemic small cell carcinoma, granulocytic sarcoma as well as undifferentiated epithelial carcinoma or metastases [8].

The immunohistochemical study allows to distinguish between these different diagnoses. Tumour cells usually express B cell markers (CD20, CD19, CD22 and CD79a), Bcl2 is expressed in 30-50% of cases. The degree of proliferation is assessed by KI67. Epstein-Barr virus (EBV) proteins can be seen in 13% of large cell B-cell lymphomas Molecularly, diffuse large-cell B lymphoma is marked´ by rearrangement of immunoglobulin heavy and light chains, translocation of the Bcl2 gene and t(14,18) in 20-30% of cases. The rearrangement of the MYC gene is unusual [8].

Lymphomas are treated with chemotherapy, using a regimen based on the cytologic type of the NHL. In the literature, the maximal surgical excision of the tumor does not seem to be associated with a better prognosis. On the other hand, the combination of biopsies and well-adapted chemotherapy seems to be the pertinent choice in the management of these patients. In the presence of such clinical and radiologic signs, the surgeon and the pathologist should consider the diagnosis of an ovarian lymphoma to determine the optimal treatment, where mutilating surgery could be avoided, while preserving the quality of life of these patients. In the presence of an ovarian tumor, the criteria of malignant lymphoma must be looked for before surgery is envisaged, as the treatment of choice may consist in biopsies and adapted chemotherapy [6].

A CHOP regimen (cyclophosphamide, adriamycin, vincristin, prednisone), with or without interferon for the less aggressive NHL, is usually administered, but a multiple-drug chemotherapy (including adriamycin, cyclophosphamide, vincristin, bleomycin, corticoids, methotrexate, and VP16) is sometimes associated with intrathecal prophylaxis [9].

The difference between primary and secondary ovarian lymphomas is important in terms of prognosis. Contrary to POL, 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. The survival seems to be shorter in ovarian NHL than in other sites. Many authors, however, report cases of POL with disease-free survival of more than 11 years [10-12].

CONCLUSION

In conclusion, an extranodal BL presenting with signs and symptoms mimicking a primary gynecologic tumor is extremely rare. In this article, we present a case of BL mimicking a gynecologic malignancy. Although extranodal BL in ovaries is a rare condition and should be included in the differential diagnosis of pelvic gynecologic tumors.

CONFLICTS OF INTEREST

None of the authors declare any conflicts of interest.

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