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Case Report

Bilateral B - cell non - Hodgkin ovarian lymphoma: A case presentation

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Abstract

The correlation between non - Hodgkin lymphoma (NHL) and the genital tract is unquestionable. The ovaries represent the most common anatomic sites. In many cases the clinical spectrum seems identical and misleads the treatment management. The prognosis is often poor due to delayed or inaccurate diagnosis. We present a case of a 63 - year - old female patient with NHL mimicking ovarian carcinoma, early diagnosed and successfully treated.

Keywords: Non - Hodgkin lymphoma; ovarian carcinoma; chemotherapy

Lymphomas mimicking ovarian tumors are uncommon and may occur as de novo or secondary as a part of systemic disease[1]. The difference between primary and secondary lymphomas is extremely important due to differences in the treatment of choice. Prognostic factors include tumor size, staging, and histological type. The most common histological types of non - Hodgkin lymphoma (NHL) are Burkitt lymphoma and diffuse large B - cell lymphoma[2]. Among them, the B - cell type is correlated with longer survival and better life quality[3]. Ovarian lymphoma may occur at any age, with a median age of 40 years[4,5]. The clinical and imaging establishment of the primary ovarian lymphoma remains controversial. Fox et al, suggested three criteria regarding the diagnosis of primary ovarian lymphoma[6]. Specifically, the tumor must be confined to the ovary regional lymph nodes or adjunctive organs at the time of the diagnosis, the bone marrow and peripheral blood should not contain any abnormal cells, and in case of appearance of extra - ovarian lesion, there must be a few months' time interval between the occurence of ovarian and extra ovarian lesions. The survival rate of primary ovarian NHL ranges from 0 - 36%[7]. The average prognosis is poor due to inaccurate or delayed diagnosis. Bilateral involvement, rapid progression of the lesion, advanced stage and signs of systematic disease indicate the severity and poor prognosis of the lesion[8].

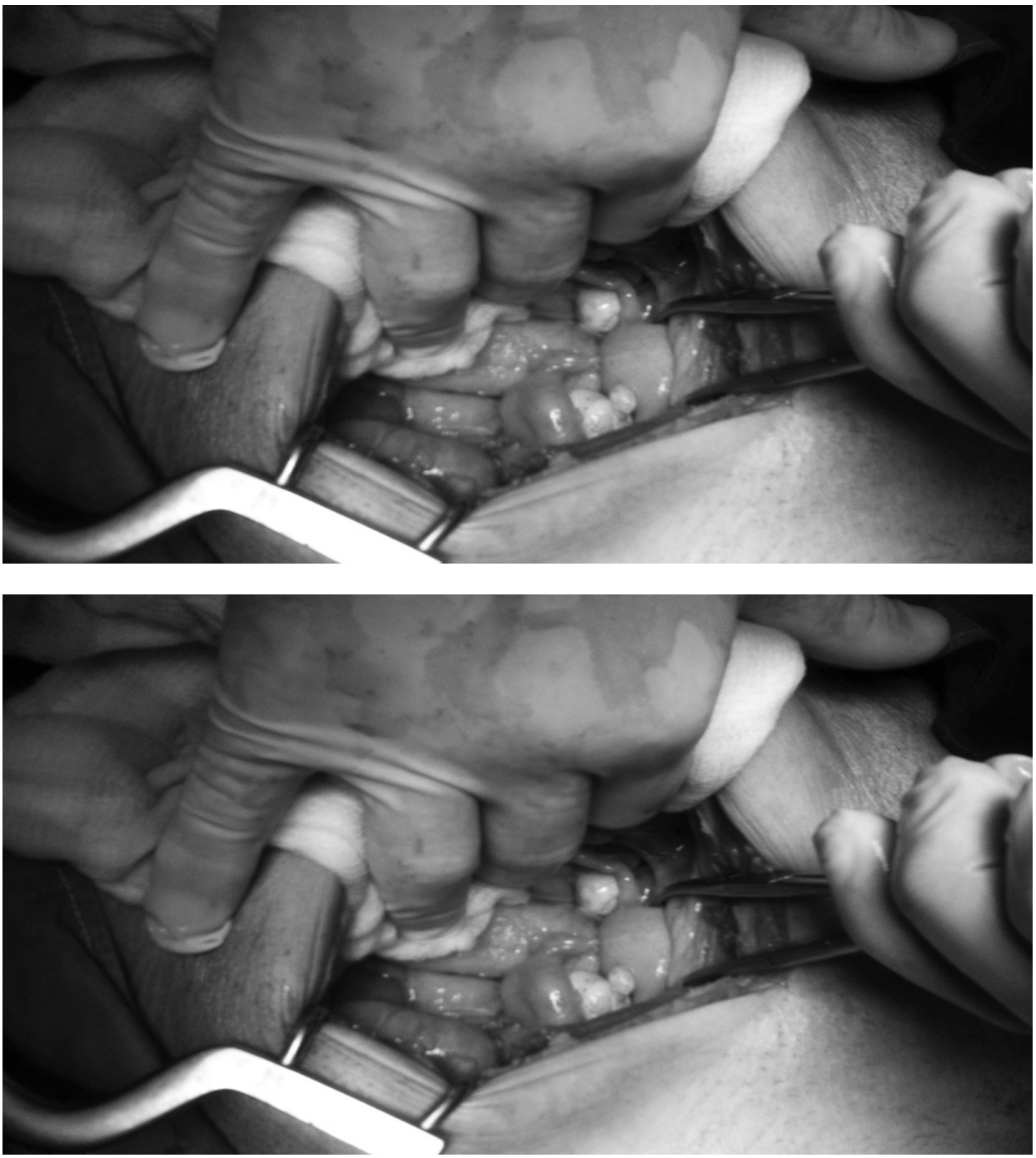
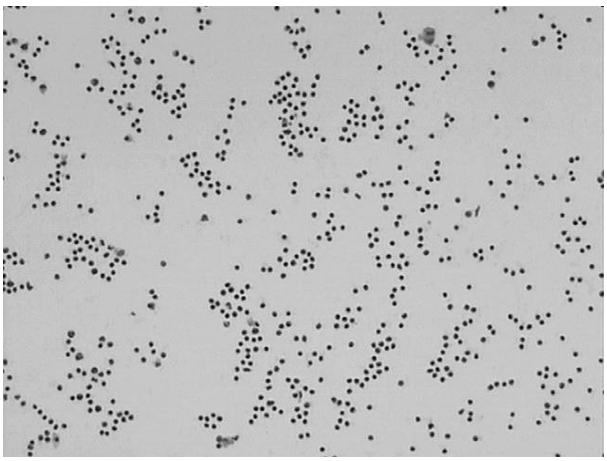


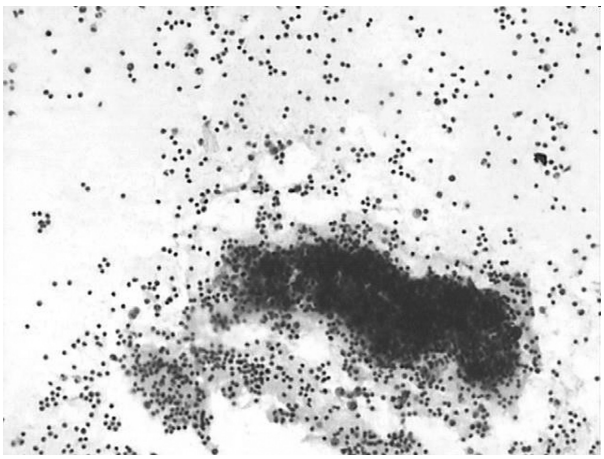
Figure 1. a) Open biopsy of the right fallopian tube, b) tumor infiltrations on the right adnexa

Case Report

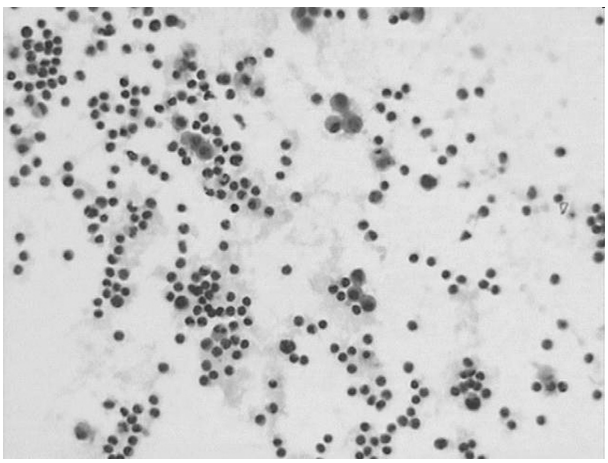
We present a case of a 63 - year - old female patient (parity 1, gravida 3) admitted to our department, complaining of distension, abdominal pain, and episodes of tinnitus during the last two months. A brain magnetic resonance imaging (MRI) was performed. The change in signal intensity of bone marrow, the presence of exoskeletal metabolism and the node -



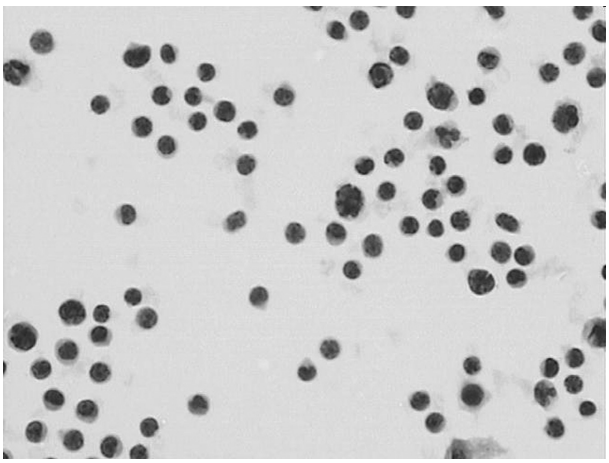
a) Papanicolaou stain X 10



b) Papanicolaou stain X 10



c) Papanicolaou stain X 20



d) Papanicolaou stain X40

Figure 2. Cytological smears from peritoneal washing dominated by numerous lymphoid cells, lying singly (a) and in clusters (b), some of them with atypical features, suggesting the diagnosis of lymphoma (c,d).

like foci in the subcutaneous tissue, were compatible with an hematological - like lymphoma lesion. Computed tomography (CT) of the abdomen revealed the presence of diffuse lymphatic blocks inside the whole retroperitoneal space and free fluid inside the peritoneal cavity, whereas both adnexa were not clearly defined. The picture was compatible with an ovarian neoplastic lesion and lymphoma. Cancer antigen 125 (CA - 125) tumor marker was 764 U/ml (normal range <36 U/ml). Transvaginal ultrasound revealed the presence of several solid, highly vascularised, riveted masses at both ovaries (159 cm^3 and 78,5 cm^3 in the left and the right ovary, respectively) and infiltrated iliac and paraaortic lymph nodes. Findings were compatible with an ovarian tumor. The gastroscopy and the colonoscopy revealed no signs of malignancy. MRI of the abdomen revealed extended infiltrated lymphatic block inside the paraaortic space, along the iliac vessels. The iliopsoas muscle, the presacral space and the parametrium were not clearly anatomically defined. Presence of free fluid inside the pleural and peritoneal cavity was noted. Papanicolaou smear was negative. Due to the large quantity of free fluid inside the pleu-

ral cavity, a pleural catheter was inserted and 1,200 ml of pleural fluid were aspirated. The cytological report revealed the presence of mesothelial cells with reactive changes, histiocytes and many lymphocytes, giving a picture of diffuse lymphatic domination. The patient underwent exploratory laparotomy and open biopsy from both fallopian tubes and the omentum. Large quantity of peritoneal fluid was drained out. There were several enlarged lymph nodes in the retroperitoneal space. The cytological examination revealed middle and large cytological elements, while the immunohistochemical examination reported the presence of B - cells, cluster of differentiation (CD)20 (+), CD79a (+), CD10 (+), B - cell lymphoma 6 (bcl - 6) protein (+), CD23(+), bcl - 2 protein (-), multiple myeloma oncogene 1 (MUM - 1) (-), CD5 (-), and a low percentage of T lymphocytes.

Overall, there was infiltration with high malignancy B - cell NHL accompanied with enlarged cells (diffuse large B - cell lymphoma - not otherwise specified, DLBCL - NOS), Epstein - Barr virus (EBV) (-) and a phenotype indicative of germinal cell origin. The bone biopsy revealed hematopoietic cell hyperplasia, increase of the megakaryocytes and limited lymphocyte presence. All these characteristics suggested the diagnosis of an extramedullary lymphoma.

The patient underwent eight cycles of chemotherapy with rituximab, cyclophosphamide, hydroxydaunorubicin, vincristine, and prednisolone (R - CHOP). The new abdominal CT showed disappearance of the pre - existing tissue between the intestinal helices and the abdominal cavity, and clear improvement of the retroperitoneal lymphatic block. The patient was followed up with CT and tumor markers every six months and was in good clinical condition one year later.

Discussion

Lymphomas mimicking ovarian tumors represent a rare entity, occurring as a regional or a systematic disease. Many studies have been conducted to establish a proper and an accurate diagnosis[9]. The management of the primary ovarian lymphoma remains controversial and the therapeutic options include surgery, chemotherapy and radiotherapy[10]. The chemotherapy protocol, regarding diffuse large B - cell lymphomas, consists of the standard R - CHOP regimen. CHOP is the acronym for a chemotherapy regimen used in the treatment of NHL: cyclophosphamide, an alkylating agent which damages DNA by binding to it and causing the formation of cross - links, hydroxydaunorubicin (also called doxorubicin or adriamycin), an intercalating agent which damages DNA by inserting itself between DNA bases, oncovin (vincristine), which prevents cells from duplicating by binding to the protein tubulin, and the corticosteroids prednisone or prednisolone. The above regimen can also be combined with the monoclonal antibody rituximab (R - CHOP). In patients with a history of cardiovascular disease, doxorubicin (which is cardiotoxic) is often omitted. The combination is then referred as COP (cyclophosphamide, oncovin, and prednisone or prednisolone) or CVP (cyclophosphamide, vincristine, and prednisone or prednisolone). The main advantage of rituximab is its essential role in the treatment management of CD20 (+) B - cell lymphoma.

Conclusion

Ovarian lymphomas are rare entities with specific treatment protocols. The physical examination and preoperative imaging findings help the diagnostic differentiation between primary and secondary lesion, whereas a multidisciplinary approach is the key to the treatment of the disease.

Conflict of interest

All authors declare no conflict of interest.

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