Case Report

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A Case of Ovary Diffuse Large B-cell Lymphoma

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Abstract

Ovarian diffuse large B-cell lymphoma is rare, and this article introduces a female patient who presents with ovarian tumor as the starting symptom, who was diagnosed with ovarian cancer during preoperative examination, and the postoperative pathological results suggested diffuse large B-cell lymphoma. This report highlights the clinical pathologic characteristics and differential diagnosis of this unusual case.

Keywords: Diffuse large B-cell lymphoma; Ovary tumor; Non-Hodgkin's lymphoma.

Case presentation

The patient was a 66-year-old woman admitted to the hospital with abdominal pain for 4 days. She had persistent vague pain in the abdomen with slight abdominal distension. She had regular menstruation and menopause at the age of 43, and had 6 pregnancies and 5 deliveries. The patient had no other past medical history.

Physical examination: No enlargement of superficial lymph nodes, abdominal compressions are slightly painful. **Gynecologic examination:** old perineal laceration of II°, senile changes of vulva, no ulceration, no superfluous growths, and the vagina is unobstructed. The uterus was normal in size, with good mobility. A 7 cm diameter mass was found in both adnexa. Both ovarian fallopian tubes have palpable masses of about 7 cm in diameter, and the surface of the right mass is nodular, movable, and mildly tender.

Laboratory examination: Ultrasound in obstetrics and gynecology, $5.6 \times 4.7 \times 4.9$ cm and $7.0 \times 3.9 \times 5.2$ cm solid hypoechoic masses were seen in the left and right adnexal areas, respectively,

with irregular morphology and internal blood flow signal. Barium enema colonography, localized stenosis in the proximal part of the ascending colon, considering the possibility of tumor invasion. **The whole abdomen CT showed:** The mass is in the bilateral adnexal area, right lower abdomen or retroperitoneal masses, bilateral multiple enlarged lymph nodes, thickening of the omentum around the right colon, considering: malignant tumor in the adnexal area, the rest are multiple metastases; malignant mesenchymal tumor in the right lower abdomen, the rest are metastases. **Hematology:** white blood cells are normal (6.42 × 10⁹ /L), Hemoglobin is lowered (114.0 g/L), elevated platelet count (305 × 10⁹/L), decreased lymphocyte percentage (0.17). Elevated CA125 (54.7U/ml). **Ascites:** Malignant tumor cells.

Surgical treatment seen in the abdominal cavity: about 50 ml of clear ascites, the bilateral ovaries were solidly enlarged, grayish white, the left ovarian mass was about 7 cm, the right ovarian mass was about 5 cm, the solid mass protruded retroperitoneally, adhered to the ipsilateral pelvic wall and uterus. A solid mass of about 6 × 6 cm in size was seen in the mesentery near the root of the colon, which was tough with clear borders and did not

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involve the intestinal lumen. The frozen section showed a malignant ovarian mass tumor. Postoperative pathology showed diffuse large B-cell lymphoma of the ovary, non-specific (germinal center type), tumor involvement in the bilateral parametrium, bilateral paracervical, bilateral fallopian tubes, vaginal stump and cervical canal. **Immunohistochemistry showed:** CD20(++); CD3(-); Ki-67(80%+); CD56(-); CD99(-); CgA(-); CK(-); CK7(-); EMA(-); Inhibin-a(-); SALL4(-); Syn(-); Vimentin(-); WT-1(-) (Figure 1) (Small mesentery). Diffuse large B-cell lymphoma, germinal center origin, immunohistochemistry showed: CD20(+); CD79a(+); PAX-5(+); CD5(-); CD3(-); CD30(-); CD38(partial +); CD10(+); MUM1(+); Bcl-6(+); P53(+); ALk(-); CD15(-); CK(-); cyclinD1(-); EMA(-); GranzymeB(-); TIA-1(-); c-Myc(-); Ki-67(80%+) (Figure 2). Tumor was detected in mesenteric lymph nodes (10/10).

The Final diagnosis was Stage IV diffuse large B-cell lymphoma of the ovary. Follow-up treatment was recommended to the oncology department. She was discharged 28 days after surgery, but was readmitted the hospital due to hypoproteinemia and infected surgical incision. The patient was cachectic, with multiple complications including cancer cachectic, secondary pernicious anemia, ureteral fistula, intestinal fistula, and poor healing of the vaginal stump. A multidisciplinary consultation recommended palliative treatment. But the patient's family abandoned the treatment and she died 3 months after surgery.

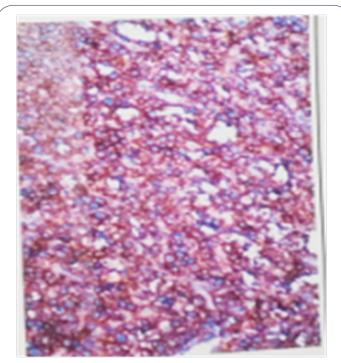


Figure 1: Ovary (HE X10).

Discussion

Etiology and incidence

Lymphoma is a group of malignant tumors arising from lymph nodes and extra-nodal lymphatic tissue [1]. It is divided into Hodgkin's lymphoma and non-Hodgkin's lymphoma. Which non-Hodgkin's lymphoma is a common type, and it can involve the ovaries, uterus, cervix, vagina and vulva in the reproductive system. The

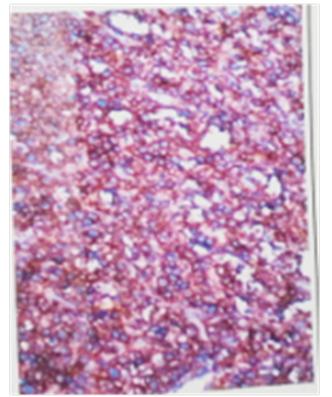


Figure 2: Small intestine (HE X10).

incidence of ovarian involvement is approximately 7%-30% [2]. Most clinical show with pelvic masses and abdominal pain. The most common type is Diffuse Large B-cell Lymphoma (DLBCL), and the most DLBCL is the germinal center type [3]. Since the ovaries is not contain lymphoid tissue, the ovarian-derived lymphoma is controversial. Most think that the tumor may originate from lymphocytes around the vascular of hepatic hilum. A few lymphocytes within the epithelium of teratoma and lymphocytes near the corpus luteum, and originates from follicular central cells [1,4]. It has also suggested that reactive lymphocytes are involved in the inflammatory response of the reproductive system, allowing them to transform into malignant tumor cells [5].

Diagnosis

DLBCL occurring in the ovary accounts for about 0.5% of non-Hodgkin's lymphomas and 1.5% of ovarian tumors, Most are secondary to generalized lymphoma, and primary disease is rare [6]. Lymphoma of the ovary has no typical clinical features, and it is difficult to distinguish it from ovarian cancer by preoperative frozen section pathology [7-9]. Which is often diagnosis according to the Postoperative pathology and immunohistochemistry. The commonly used indicators in DLBCL immunohistochemistry including [5] CD20, CD3, CD5, CD10, Bcl-6, Ki-67, MYC. BCL2 and (or) BCL6 rearrangements, often called Double-hit Lymphoma, also has a poor prognosis [19]. Syn and CD99 negativity can be distinguished from Peripheral primitive neuroectoderal tumor. CK(-) can be distinguished from tumors of Epithelial neoplasms. In this case, Ki67 >80% positive, this indicates that tumor cells are actively proliferating and poor prognosis [10] However, it has also been suggested that survival is not related to Ki-67 expression [11]. The differential diagnosis between secondary and primary lymphoma which from ovary is difficult, but this is important for the

assessment of treatment and prognosis [14]. Primary tumors are mostly confined to the ovary, while secondary tumors are mostly considered to be systemic disease of DLBCL involving the ovary [12]. Secondary ovarian DLBCL can be divided into two types, (1) The early stages manifest as unknown extra ovarian disease; (2) secondary ovarian involvement in disseminated systemic DLBCL [13]. To diagnose primary lymphoma, systemic lymphatic disease must be excluded, and early primary ovarian lymphoma usually has a good prognosis [7]. It has been suggested that MRI of bilateral ovarian masses is a clue to diagnosis of ovarian lymphoma, and once the disease has spread to the abdomen, the differential diagnosis between primary and secondary is more difficult [12]. The diagnostic criteria for the diagnosis of primary ovarian lymphoma which proposed by Fox and Langley that most people use today is that [15], 1) tumor confined to the regional lymph nodes or accessory organs of the ovary at the time of diagnosis; 2) absence of abnormal cells in the bone marrow or peripheral blood; 3) any extra-ovarian disease must occur within a few months of the appearance of the ovarian lesion.

In this case, the patient had a normal temperature, no night sweat, and multiple extra-nodal organ invasions before surgery. And belonged to stage IV type. At the time of diagnosis, the patient had lymph node involvement in the ovary, and mesentery involvement with regional lymph node metastases. The patient's condition changes, and no bone marrow smear is done to determine whether there are abnormal cells in the bone marrow. Therefore, the origin of the disease cannot be clarified.

Treatment

Based on the difficulty of preoperative, it has been suggested that if intraoperative frozen section is highly suspicious that tumors from the reproductive system, surgeons should continue the cytoreduction, otherwise further treatment options should be choose by the final pathological diagnosis [12]. Rituximab in combination with CHOP scheme for Non-Hodgkin is an effective treatment [16]. But, a combination of surgery-based treatment with radiotherapy is currently used, which is inconsistent with the recommended treatment for DLBCL, and the effect of surgery combined with chemo radiotherapy on patient prognosis is unclear. Most believe that the combination of surgery and chemotherapy can prolong the survival of patients [17].

Prognosis

The prognosis of DLBCL is closely related to the age, clinical stage, lesion extent, histology, systemic symptoms, and lactate dehydrogenase [12,18]. The patient is older, multiple extra-junctional lesions, these all suggested a poor prognosis. Death due to multiple organ infections such as intestinal fistula, ureter stoma and hypoproteinemia. Two other cases of ovarian DLBCL in our gynecology department within a decade. Both of them died within six months after surgery due to periprocedural complications. Combined with this case, we should strengthen perioperative management so that they can receive chemotherapy.

Conclusion

Diffuse large B-cell lymphoma arising in the ovary is rare, The incidence of DLBCL in the ovary is low. Most of them are diagnosed by postoperative histopathology, and there are few cases who only receive chemotherapy, and it is difficult to understand

the impact of surgery on the prognosis of patients. We believe that it is necessary to collect more data in the clinic. For patients with high clinical suspicion of ovarian DLBCL, the therapeutic value of surgical treatment for the disease is fully explored, and a more reasonable and perfect comprehensive treatment method is sought to prolong the survival of patients.

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