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Case Report

Primary Ovarian Lymphoma in Late Pregnancy: A Case Report

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Abstract

Involvement of the ovary with malignant lymphoma is a well-known late manifestation of disseminated nodal disease. Primary ovarian Burkitt lymphoma is very rare and mainly affects young children. We present a case of a 25-year old woman at 37 weeks pregnant with sporadic Burkitt lymphoma who presented as having ovarian cancer. The patient was managed via elective cesarean section and unilateral oophorectomy. Histopathological examination and immunohistochemical stains were carried out, revealing non-Hodgkin B-cell Burkitt lymphoma. After the diagnosis, the patient was referred to the oncology center for chemotherapy.

Keywords: Extranodal lymphoma, Burkitt lymphoma, ovarian tumor, pregnancy.

INTRODUCTION

Primary ovarian non-Hodgkin lymphoma accounts for 0.5% of all non-Hodgkin lymphomas and 1.5% of all ovarian neoplasms. Since there is no lymphatic drainage for the ovaries, primary malignant lymphomas of the ovary are extremely rare.

In 2008, Nelson *et al*. [1] reported six cases of secondary ovarian lymphoma encountered at the Mayo Clinic over a period of 20 years. In 2007, Woodruff *et al*. [2] reported 35 cases of secondary ovarian lymphomas that were recorded in the ovarian tumor registry of the American Gynecological Society.

CASE REPORT

A 25-year-old woman (gravida 2, para 1) was admitted to our hospital with increasing abdominal volume at her final weeks of gestation. Physical examination revealed that the uterine fundus was reaching up to the epigastrium. Ultrasound revealed a huge mass filling the posterior pouch and extending to the left maternal side, the size of which could not be measured because it extended beyond the ultrasound field. Heterogeneous internal echoes were observed. Hypervascularity was present on color Doppler imaging.

MRI scan revealed normal fetus (Figure 1A) along with a large maternal pelvic mass located posterior to the uterus and to the right which elicited a signal isointense relative to the myometrium, with high T2 signal areas and a central signal void vessel. It measured about 13.3 × 17.7 × 20 cm in size along the anteroposterior aspect (Figure 1B). It was compressing the pelvic structures, including the lower uterine segment. Multiple enlarged amalgamated retroperitoneal lymph nodes of varying size were seen encasing the aorta and renal vessels. A lower cesarean section was done through a large lower abdominal vertical incision to deliver the baby and reveal the large pelvic mass replacing the whole right ovary. The left ovary was not visualized. The right ovarian mass was excised and sent to our histopathology department. Gross examination revealed a single mass which was oval in shape, greyish tan in color, soft to firm in consistency, and measured 23 × 17 × 8 cm in size. The cut section was homogenous and nodular, with areas of hemorrhage. No necrosis or cystic changes could be seen (Figure 2). Microscopic examination revealed total replacement of ovarian tissue by diffuse sheets of monotonous, medium-sized cells, with round nuclei, distinctly clumped chromatin, multiple nucleoli, and dense amphophilic, vacuolated cytoplasm with brisk mitotic activity and typical low-power (starry sky)

appearance, with tingible body macrophages (Figure 3,4). Minute residual ovarian tissue was noted. Immunohistochemical profiling revealed that the malignant cells were diffuse positive for CD45, CD20, CD10, C-myc (Figure 5), and Ki67 (almost 100%), with focal positivity for CD3 and Bcl-6. The malignant cells were negative for CD5, CD23, Cyclin D1, Alk-1, AFP, OCT-4, CD117, D2-40, PLAP, EMA, and BCl-2. Pathologic assessments were consistent with non-Hodgkin lymphoma, with morphological and immunohistochemical features indicating B-cell type Burkitt lymphoma (BL).

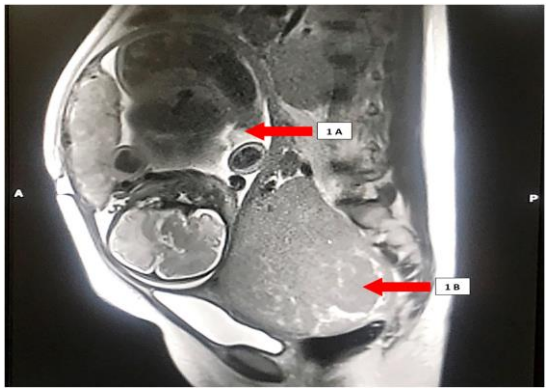

Fig-1A: Fetus, Fig-1B: Tumor

Fig-2: Gross Photo showing a single bisected mass, measuring 23 × 17 × 8 cm in size; the cut section is homogenous and nodular, with areas of hemorrhage.

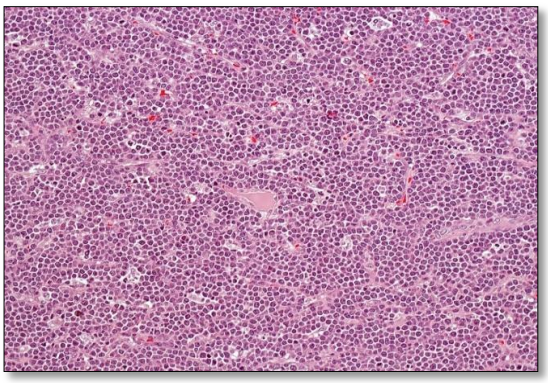

Fig-4: Histopathology examination demonstrates round nuclei, distinctly clumped chromatin, multiple nucleoli, and dense amphophilic, vacuolated cytoplasm with brisk mitotic activity (H&E staining; ×40).

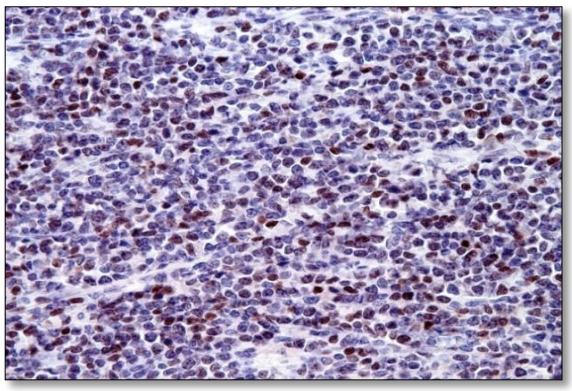

Fig-5: Positive c-MYC (immunostaining; ×40).

DISCUSSION

The majority of lymphomas involving the ovary are of B-cell phenotype; BL and diffuse large B-cell lymphoma are the most common variants, with BL usually presenting bilaterally [3]. Intra-abdominal BL is often diagnosed at a very late stage due to nonspecific symptoms, usually presenting with abdominal pain, nausea, vomiting, amenorrhea, and abdominal distension [4]. Around 70% of BL patients present with head and neck involvement at an advanced stage of the illness [5]. BL is a highly aggressive mature B-cell lymphoid malignancy, first described by a British surgeon (Denis Parsons Burkitt) in Africa. It demonstrates translocation between the c-MYC gene and immunoglobulin heavy locus, most commonly t(8;14)(q24;q32). The illness has three variants: endemic, sporadic, and immunodeficiency-associated. The endemic variant is strongly connected with Epstein–Barr virus infection and is diagnosed more often in endemic malaria regions [6]. It usually involves the jaw or facial bones and spreads to the bone marrow, peripheral blood, meninges, testes, ovaries, kidneys, and breasts. Sporadic BL is mostly diagnosed in young adults in the United States and Western Europe. It presents as an extranodal manifestation, usually in the abdominal cavity [7]. Immunodeficiency-associated BL is mostly observed in HIV-positive patients, though it can also be seen in patient's post-organ transplantation or patients with primary immunodeficiency [7]. The hypothesis for the origin of primary ovarian lymphoma (POL) suggests the presence of pre-existing lymphoid tissue in the ovary [8]. Although lymphocytes are absent in the ovaries, those surrounding the blood vessels at the hilum or related to the corpus luteum may be the origin for the tumor cells [9]. Some pieces of literature claim that reactive lymphocytes may be involved with the ovary in patients with pelvic inflammatory disease or endometriosis, which can transform into POL. BL consists of monomorphic, medium-sized cells with basophilic cytoplasm, a high proliferation fraction, and a 'starry sky' pattern [10]. The criteria presented by Fox *et al*. [11] for diagnosing POL are as follows: (a) The lymphoma is confined to the ovary or the adjacent lymph nodes or structures at

diagnosis; (b) There is no evidence of the disease in the blood or bone marrow; (c) The lymphomateous lesions occur at non-ovarian sites and appear at least a few months after ovarian involvement [11].

The differential diagnosis of malignant ovarian lymphoma must include poorly differentiated ovarian carcinoma and ovarian metastasis, granulosa cell tumors, hypercalcaemic small-cell ovarian carcinoma, or dysgerminoma [12].

CONCLUSION

Primary ovarian BL is a rare condition which is included in the category of non-Hodgkin lymphoma. Patients suffering from ovarian BL commonly undergo surgery after the ovarian tumor has been detected, but surgical treatment is not the treatment of choice: an early multi-agent chemotherapy protocol improves the prognosis and the survival rate.

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