An Appendix Origin Burkitt's Lymphoma Spreading into Genital Tract

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ABSTRACT

Burkitt's lymphoma (BL) occurs commonly in children and young adults, whereas bilateral ovarian involvement is extremely rare. We report a patient with unusual symptoms for BL, mimicking an ovarian neoplasm. Here, our aim was also to point out that BL can spread into the genital tract.

A 60-year-old woman presented with the complaint of abdominal pain for a few months. Abdominal distension and sensibility were observed upon physical examination of the patient, but no common "B" symptoms associated with lymphoma were noted. Our imaging studies revealed bilateral adnexal masses suggesting an ovarian tumour with positive laboratory findings. We performed an exploratory laparotomy and a total hysterectomy, bilateral salpingo-oophorectomy and right hemicolectomy with appendectomy. The hemicolectomy process was needed to reduce the tumour burden and because of the risk of intestinal obstruction risk that could arise from the tumour mass in the ileocecal valve. Observation of a frozen specimen indicated BL, so the surgery was ended.

Although BL involving the genital tract is a rare condition in patients who have pelvic masses, BL can be one of the diseases that should be considered for specific diagnosis.

Keywords: Burkitt's Lymphoma, Abdominal Pain, Genital Tract

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Introduction

Burkitt's Lymphoma (BL) is one of the Non- Hodgkin Lymphomas that have high grade and aggressive B-cells. It was first identified in Africa in 1956 by an Irish surgeon, Denis Parsons Burkitt. In 1958, this disease was then announced worldwide with this surgeon's name.^{1,2}

BL is a malignancy that is commonly seen in children and young adults. In this group of patients, BL rarely involves the ovaries and genital organs, although it can be diagnosed by ultrasonography, which shows an increase in the dimensions of the ovary. In 26% of the cases, ovarian involvement is only found only during autopsy.^{3,4}

While primary lymphoma of the ovary is rarely seen, 1.5% of ovarian tumours and 0.5% of Non-Hodgkin lymphomas are primary ovarian lymphomas.^{5,6,7} The symptoms of BL that

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mimic primary gynaecological cancer are often non- specific, and it can be diagnosed as abdominal distension and abdominal pain.⁸ In some cases, "B" symptoms, including fever, weight loss, night sweats and fatigue, which are associated with lymphomas, can be determined.⁹ In this case report, the patient with BL having a primary appendix origin did not show "B" symptoms but only abdominal pain, indicating genital tract involvement. In this report, our aim was also to point out that BL can spread into the genital tract.

Case Report

The patient, aged 60, was G6, P3 and had presented at our clinic with a complaint of abdominal pain that had been ongoing for a few months. Abdominal distension and sensitivity were observed during physical examination of the patient. The dimensions of the uterus were determined as 11x9x5 cm, while endometrial echo was evaluated as 4 mm by a trans vaginal ultrasonographic observation. Additionally, the left adnexal region contained a solid 6x5 cm mass with disordered boundaries and the right adnexal region contained a 5cm semisolid cystic mass, also with disordered boundaries. Minimal free body liquid was seen in Douglas. Tumour marker laboratory evaluation revealed a CA125 level of 179.8 U/ mL, while the CA19-9 level was 36.1 U/mL. The other laboratory tests that contained routine biochemical and hemogram tests were also normal.

An exploratory laparotomy was performed. The intra op-

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erative evaluation indicated that the uterus was larger than normal and fragile. Tumoral implants were also seen in the bilateral ovaries and uterine tubes. The ileocecal valve contained a tumoral mass of approximately 6 cm that originated from the appendix. Focal tumours were also implanted throughout the whole colon as well as in the omental cake. Abdominal hysterectomy + bilateral salpingo -oophorectomy + total omentectomy and right hemicolectomy + appendectomy + side to side anastomosis were performed.

A frozen tissue observation showed histopathological findings that were consistent with a lymphoid tumour. Macroscopically, the examination of hysterectomy and salpingo- oopherectomy materials had the appearance of fresh fish (Figure 1).



Figure 1 : The macroscopic examination of hysterectomy and salpingo- oopherectomy materials

Histopathological examination revealed diffuse lymphoid tumoral infiltration (Figure 2). Neoplastic cells were small to medium sized (Figure 3), with many mitotic figures. Neoplastic cells showed positivity for CD10 (Figure 4), BCL-6, CD20 (Figure 5) and CD79a. CD43 positivity was only seen in scattered cells. The Ki-67 proliferation index was about 95%. Histopathological and immunohistochemical findings were consistent with BL.

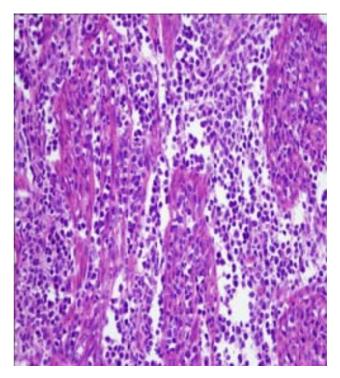


Figure 3: Non-cleaved neoplastic cells were small-medium sized (HEx400).

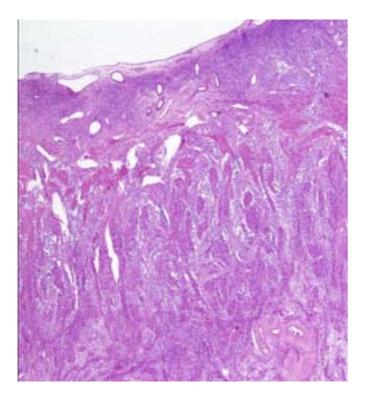


Figure 2: Lymphoid tumoral infiltration in endometrium and myometrium (HEx40).

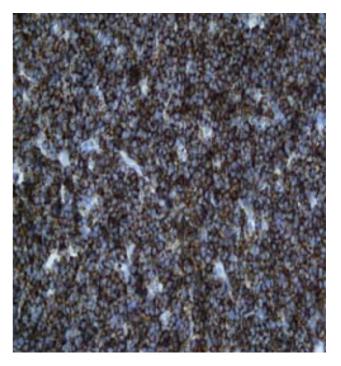


Figure 4: CD20 immunuexpression in lymphoid cells.

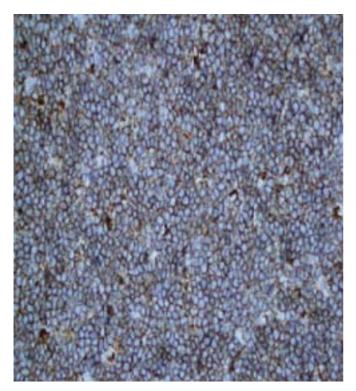


Figure 5: CD10 positivity in neoplastic cells.

After the histopathological results and diagnosis that the bulk was Burkitt's Lymphoma that had originated from the appendix and spread into the genital tract, the treatment of the patient continued with the adjuvant chemotherapy [CHOP (Cyclophosphamide, Hydroxydaunorubicin, Oncovin(vincristine) and Prednisone)].

Conclusion

Although BL frequently occurs during childhood, it can also affect the population aged between 3 and 63 years.^{9,10} While "B" symptoms associated with lymphomas, such as fever, weight loss, night sweats, or fatigue, can be seen in BL, non-specific symptoms including abdominal distension, abdominal pain, palpable pelvic masses, and pelvic pain can also be found in the patients who have gastrointestinal and genitourinary tract involvement.⁸ In our case, the main reason for the patient attending our clinic was abdominal pain that was non-specific for lymphomas rather than having "B" symptoms. For this reason, our ultrasonographic observations for pre-diagnosis were considered to possibly point to an ovarian neoplasm in this patient.

The pre-diagnosis as a primary ovarian tumour led to the total hysterectomy, bilateral salpingo-oophorectomy and right hemicolectomy with appendectomy techniques that were performed. The hemicolectomy process was performed due to the need to reduce the tumour burden and because of the risk of intestinal obstruction that could arise from the tumour mass in the ileocecal valve. As after the finding of BL after observation of frozen tissue, the pelvic and para-aortic lymphadenectomy procedures were not conducted and the operation was concluded.

Since BL consists of high grade and aggressive B- cell lymphomas, patients should get more rapid diagnosis and treatment. Early diagnosis and treatment are crucial because of the high doubling- time.¹⁰ Delays in treatment can cause fast growth of the tumour mass and increases in morbidity and also mortality.

BL has a high sensitivity to chemotherapy, so the place of surgery in the treatment, and the choice of treatment only by chemotherapy or by both surgery and chemotherapy, should be discussed.¹¹ Morbidity and mortality can increase due to surgical resection, and because of the delayed chemotherapy, the tumour mass can grow again.⁹ Because this tumour clinically imitates the primary ovarian cancers by involving the genital tract, it is difficult for clinicians to make a diagnosis and select the type of the treatment. In addition, some studies have shown success in the treatment of BL when the treatment is done by surgery combined with chemotherapy.¹²

Differentiation of BL located in the pelvic region from other malignancies is seldom possible before surgery and generally, in patients undergoing exploratory laparotomy to confirm colon or ovary neoplasms, it can appear intra operatively. In our opinion, the size of the tumoral mass is one of the key points for gynecologists to consider when making their treatment decision. Generally, big masses are removed surgically, whereas for the cases where no distinctive mass is observed, surgery remains limited. In our case, the masses were sufficiently large to make a decision to perform a surgical treatment.

In conclusion, because of the inadequacy of pre-diagnosis and the mimicking of primary ovarian cancer by BL involving the genital tract, gynecologists can be directed to surgery, as in our case. Therefore, patients who have a pelvic mass should be evaluated for BL as one of the diseases that should be considered for the specific diagnosis.

Genital Sisteme Yayılan Apendiks Orijinli Burkitt'in Lenfoması

Burkitt Lenfoma (BL) genellikle çocuk ve genç erişkinlerde görülmekte olup, bilateral ovaryan tutulum çok nadirdir. Olgumuzda, BL için alışık olunmayan ve ovaryan neoplazileri taklit eden semptomları mevcut olan hastamızı sunacağız. Bu çalışmada, amacımız BL'nin genital sisteme yayılım gösterebildiğini kanıtlamaktadır.

60 yaşında bayan hasta birkaç aydır devam eden karın ağrısı şikayeti ile kliniğimize başvurdu. Hastanın yapılan abdominal muayenesinde, abdominal distansiyon ve hassasiyet saptandı ancak lenfomayla ilişkili genel "B" semptomları gözlemlenmedi. Görüntüleme çalışmalarımız, pozitif laboratuar bulguları ile bilateral adneksiyal kitlelerin ovaryan tümürü desteklediğini göstermiştir. Eksploratris laparatomi, total histerektomi, bilateral salpingo ooferektomi ve sağ hemikolektomi ile apendektomi prosedürleri uygulandı. Hemikolektomi işlemini ileoçekal bileşkedeki kitle nedeniyle gelişebilecek olası barsak obstrüksiyon riskinden ve tümör yükünü azaltma isteğimiz dolayısıyla gerçekleştirdik. Frozen inceleme sonucu, olgunun Burkitt lenfoma olarak rapor edilmesi nedeniyle, operasyonu sonlandırdık.

Pelvik kitlesi olan hastalarda genital sistem tutulumlu BL nadir bir durum olmasına rağmen; ayırıcı tanılarda, BL, düşünülmesi gereken hastalıklardan biri olduğu akılda tutulmalıdır.

Anahtar Kelimeler: Burkitt lenfoma, Karın ağrısı, Genital sistem

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