Burkitt Lymphoma: Involvement of Both Ovaries and Kidneys in an Adult: Case Report

Burkitt Lenfoma: Her İki Yumurtalık ve Böbrek Tutulumlu Yetişkin Olgu

Murat ACAR,[a]
Servet TATLI,[b]
Peter M. SADOW,[c]
Stuart G. SILVERMAN[b]

[a]Department of Radiology,
Bezmiâlem Vakıf University
Faculty of Medicine, İstanbul
Divisions of
[b]Abdominal Imaging and Intervention,
[c]Pathology,
Harvard Medical School
Brigham and Women's Hospital,
Boston

Geliş Tarihi/*Received:* 05.04.2011
Kabul Tarihi/*Accepted:* 11.10.2011

Yazışma Adresi/*Correspondence:*
Murat ACAR
Bezmiâlem Vakif University
Faculty of Medicine,
Department of Radiology, İstanbul,
TÜRKİYE/TURKEY
drmacar@hotmail.com

doi: 10.5336/medsci.2011-24173

ABSTRACT Burkitt lymphoma is an uncommon type of non-Hodgkin lymphoma that often involves extranodal sites. The disease predominantly affects children and has an aggressive clinical course. We report the imaging findings of a 23-year-old woman with an atypical Burkitt lymphoma that involved both ovaries and kidneys. The patient has remained in complete remission three years after the initiation of treatment.

Key Words: Burkitt lymphoma; ovary; kidney

ÖZET Burkitt lenfoma non-Hodgkin lenfomanın ekstranodal tutulumuyla seyreden nadir bir şeklidir. Hastalık özellikle çocukluk çağında görülür ve agresif bir klinik seyir gösterir. Burada bilateral over ve böbrek tutulumu saptanan ve tedavi sonrası 3 yıldır tam remisyonda olan 23 yaşındaki atipik Burkitt lenfoma olgusunun görüntüleme bulguları sunuldu.

Anahtar Kelimeler: Burkitt lenfoması; over; böbrek

Turkiye Klinikleri J Med Sci 2012;32(6):1758-61

Burkitt lymphoma is a highly aggressive form of non-Hodgkin lymphoma (NHL) derived from B-lymphocytes. The disease was first described by Burkitt in 1958 as an endemic disease of central Africa.[1] Later, a nonendemic, sporadic type was described.[2] Burkitt lymphoma occurs mostly in male children with mean age of 7 years; however, it has been observed in adults.[3] The Epstein-Barr virus has been shown to be related to the more common endemic African form (up to 95% of cases) but the relationship is less clear in the nonendemic form.[4]

In the endemic form, the disease progresses rapidly and often presents in extranodal sites such as the jaw, and other facial bones, mesentery, and gonads.[1] The nonendemic form typically presents with a rapidly growing abdominal or pelvic tumor that often involves the ileum, cecum and mesentery.[5] The kidney and liver might be involved secondarily.[2] The nonendemic form Burkitt lymphoma can cause pain, nausea, vomiting, bowel obstruction,and gastrointestinal bleeding.[2] Presenting symptoms may mimic acute appendicitis or intussusception. Rarely, the ovaries are primarily in-

volved.[3] When the ovaries are involved, the weight of the tumor may cause ovarian torsion. Because of tumor hypervascularity, intratumoral hemorrhage may develop.[2] Bilateral kidney and ovary involvement has rarely been noted.[2] In this article, we report the imaging findings of an adult who presented with Burkitt lymphoma that involved both ovaries and kidneys.

CASE REPORT

A previously healthy 23-year-old woman was admitted to our hospital because of palpable bilateral adnexal masses. She had experienced increasing bilateral leg pain and anorexia. Transvaginal US revealed bilateral, heterogeneously hypoechoic, solid adnexal masses (Figure 1a). A subsequent enhanced computerized tomography (CT) scan obtained at portal venous phase revealed heterogeneously enhancing solid masses in both adnexa (right, 6.2 cm; left, 6.8 cm) with well-marginated, lobulated contours (Figure 1b). A small amount of peritoneal fluid was also present in the pelvis. The uterus was normal. In addition, there were multiple, ~2.5 x 2.0 cm, well-marginated, intraparenchymal soft tissue attenuation masses in both kidneys that showed homogeneous enhancement relative to the rest of the renal parenchyma (Figure 1c). A complete blood count demonstrated a decreased hematocrit (31.9), mildly elevated white blood count (13.2/μL), and an elevated platelet count (573,000/μL). A primary ovarian neoplasm was suspected, and the patient underwent exploratory laparotomy with bilateral salpingoophorectomy, omentectomy, and pelvic lymph node dissection.

On gross pathology, both ovaries were enlarged and lobulated in contour; cut surfaces were heterogeneous, solid, and tan/white in color. Microscopically, the ovaries were involved extensively with diffuse, sheet-like infiltrates of large atypical lymphoid cells with a high nucleus-to-cytoplasm ratio (Figure 1d). Tumor cell markers were positive for CD20 and Bcl-6. CD3, CD5 and CD43 highlighted surrounding small reactive T cells. The Ki-67 proliferative index of tumor cells was virtually 100%. There was no evidence of involvement of the lymph nodes or omentum. In situ hybridization for Epstein-Barr virus encoded small RNA was negative. The final pathologic diagnosis was high-grade, B cell type, non-Hodgkin lymphoma consistent with Burkitt lymphoma. Cytogenetics revealed a translocation of t (8:22) (q24:q11) involving the myc gene, a variant seen in Burkitt lymphomas. Bone marrow biopsy was negative for involvement.

Follow-up CT examination obtained two months after the initiation of chemotherapy showed no evidence of disease (Figure 1e). The patient has been in complete remission for three years.

DISCUSSION

The imaging findings of ovaries in our case were not typical of primary epithelial ovarian tumor. The epithelial tumors are typically cystic, and occur most commonly in elderly adults. Our patient had no history of malignancy, so it was unlikely that the adnexal masses were metastases. Imaging findings of metastatic ovarian tumors, so-called Krukenberg tumors, are variable and may present as a predominantly solid mass as in our case or a mass with mixture of cystic and solid components.[6,7] Most non-genital cancers that metastasize to the ovaries arise from gastrointestinal tract, particularly from the stomach, and may also have a predominantly solid appearance.[7] Although sex-cord stromal tumors of the ovary are solid on imaging, these tumors, particularly granulosa cell tumors, often present with excess steroid hormone production, which may result in precocious puberty or virilization.[6] In the absence of clinical signs of excessive hormone production, an ovarian sex-cord stromal tumor was also unlikely in our case. Finally, a torsioned ovary can become quite large and heterogeneous and may mimic solid ovarian neoplasm; however, torsion of both adnexa occurs extremely rare.[8]

In addition to lymphoma, differential diagnosis of multiple solid renal masses include benign primary neoplasms such as angiomylipomas and oncocytomas, malignant primary neoplasms such as renal cell carcinomas (RCC), metastases, leukemia, and non-neoplastic conditions such as pyelonephritis and infarction.[9,10] Lymphomatous

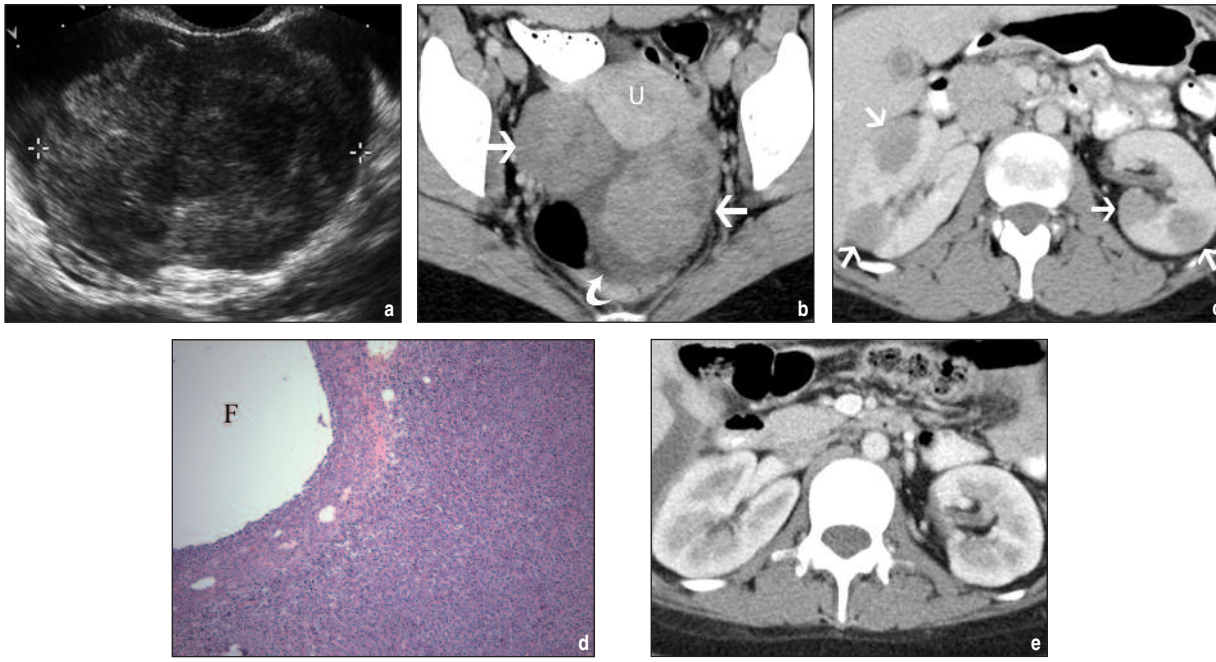

FIGURE 1a-e: 23-year-old woman with palpable pelvic masses. Ultrasonography shows **(a)** a large, 6 cm, heterogeneous, solid left ovarian mass. Contrast-enhanced computerized tomography (CT) image **(b)** shows both ovaries replaced by heterogeneously enhancing, solid masses (arrows). Note also normal looking uterus (U) and small amount of ascites (curved arrow). Contrast-enhanced CT image **(c)** demonstrates multiple, well-defined, homogeneous soft tissue attenuation masses in both kidneys (arrows). Photomicrograph of histopathologic specimen **(d)** demonstrates a population of large, atypical B cells replacing the majority of the ovarian parenchyma, which only identified by the presence of an ovarian follicle (F) (H&E stain, x 20). Follow-up CT examination **(e)** two months after the initiation of chemotherapy shows complete regression of the renal masses.
(See for colored form http://tipbilimleri.turkiyeklinikleri.com/)

involvement of the kidneys are rare; it is detected in only 3%-8% of patients undergoing CT staging for lymphoma.[9] Lymphoma typically involves the kidney in one of several recognizable patterns, including solitary or multiple renal masses, renal involvement from contiguous retroperitoneal or peritoneal disease, and diffuse renal infiltration.[9,10] Lymphomatous masses of the kidney tend to be homogeneous with mild enhancement, and exhibit little mass effect.[9] In our case, renal masses thought to be metastases. However, ovarian malignancies usually do not metastasize to the kidneys. Multiple or bilateral RCCs are typically seen in patients with genetic syndromes such as von-Hippel-Lindau and other familial RCC syndromes.[10] Pyelonephritis was also unlikely in our patient since clinical sings and symptoms were absent. Infarction was unlikely as there were no predisposing conditions such as cardiac thrombus. Both pyelonephritis and infarction may exhibit a wedge-shaped abnormality, however, pyelonephritis is typically, and often accompanied by perinephric stranding. Although most angiomyolipomas can be diagnosed using CT or MR imaging by presence of tumoral fat; multiple fat-poor angiomyolipomas with minimal fat are generally seen only in patients with tuberosclerosis.[10] Finaly, multiple oncocytomas may present with similar imaging findings but would not explain the ovarian findings.

This patient underwent oopherectomy and lost her potential for fertility. If imaging interpretation had raised the question of lymphoma, percutaneous biopsy of renal masses could have been performed and diagnosis of Burkitt lymphoma could have been made. Although surgical open biopsy still a common demand when lymphoma is suspected, image-guided biopsy has shown to be a safe and accurate method in the diagnosis and classification of lymphomas.[10,11] Addition of large needle (>18 gauge) samples to flow cytometry and cytological analysis of fine needle biopsy samples provide the highest diagnostic yield, and the most

accurate lymphoma subtype classification.[12,13] Bone marrow biopsy may also establish the diagnosis. Although it was indicated for the initial diagnosis and for resection of the disease in the past, laparotomy is not recommended in current guidelines. Chemotherapy is the mainstay of treatment and surgery or radiation therapy has no role in the treatment of Burkitt lymphoma.[3,13]

In conclusion, bilateral, multiple, homogenously enhancing solid renal masses in the presence of bilateral ovarian masses should raise the suspicion for lymphoma, especially in children or young patients. Imaging interpretation and recommendations should be done carefully so that these patients should undergo lymphoma workup before any surgical excision is undertaken.

REFERENCES

1. Burkitt D. A sarcoma involving the jaws in African children. Br J Surg 1958;46(197):218-23.
2. Lu SC, Shen WL, Cheng YM, Chou CY, Kuo PL. Burkitt's lymphoma mimicking a primary gynecologic tumor. Taiwan J Obstet Gynecol 2006;45(2):162-6.
3. Watmore AE, Murray JA, Potter AM. Nonendemic Burkitt's lymphoma with complex chromosome abnormalities involving chromosomes 2 and 8. Cancer Genet Cytogenet 1985;16(4):283-8.
4. Baloglu H, Turken O, Tutuncu L, Kizilkaya E. 24-year-old female with amenorhea: bilateral primary ovarian Burkitt lymphoma. Gynecol Oncol 2003;91(2):449-51.
5. Brady G, MacArthur GJ, Farrell PJ. Epstein-Barr virus and Burkitt lymphoma. J Clin Pathol 2007;60(12):1397-402.
6. Jung SE, Rha SE, Lee JM, Park SY, Oh SN, Cho KS, et al. CT and MRI findings of sex cord-stromal tumor of the ovary. AJR Am J Roentgenol 2005;185(1):207-15.
7. Choi HJ, Lee JH, Kang S, Seo SS, Choi JI, Lee S, et al. Contrast-enhanced CT for differentiation of ovarian metastasis from gastrointestinal tract cancer: stomach cancer versus colon cancer. AJR Am J Roentgenol 2006;187(3):741-5.
8. Chang HC, Bhatt S, Dogra VS. Pearls and pitfalls in diagnosis of ovarian torsion. Radiographics 2008;28(5):1355-68.
9. Urban BA, Fishman EK. Renal lymphoma: CT patterns with emphasis on helical CT. Radiographics 2000;20(1):197-212.
10. Bosniak MA. The small (less than or equal to 3.0 cm) renal parenchymal tumor: detection, diagnosis, and controversies. Radiology 1991;179(2):307-17.
11. Silverman SG, Lee BY, Mueller PR, Cibas ES, Seltzer SE. Impact of positive findings at image-guided biopsy of lymphoma on patient care: evaluation of clinical history, needle size, and pathologic findings on biopsy performance. Radiology 1994;190(3):759-64.
12. Hesselmann V, Zähringer M, Krug B, Wesselmann C, Haferkamp K, Wickenhauser C, et al. Computed-tomography-guided percutaneous core needle biopsies of suspected malignant lymphomas: impact of biopsy, lesion, and patient parameters on diagnostic yield. Acta Radiol 2004;45(6):641-5.
13. Güner H, Onan MA, Kurdoğlu M, Tıraş MB, Erdem Ö. Case of primary ovarian Burkitt lymphoma. Turkiye Klinikleri J Gynecol Obst 2005;15(2):103-4.