Case Report

Burkitt Lymphoma Presenting as Menorrhagia and a Vaginal Mass in an Adolescent

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A B S T R A C T

Background: Menorrhagia is a common gynecologic complaint among adolescents, which rarely is secondary to malignancy. Burkitt lymphoma can mimic gynecologic malignancy, however it is rarely seen in adolescents. Burkitt lymphoma of the gynecologic tract requires early diagnosis and intervention for optimal outcomes.
Case: We report a case of a 15-year-old adolescent who had multiple admissions for menorrhagia that was thought to be secondary to anovulatory bleeding until pelvic ultrasound revealed a large 8-cm vaginal/cervical mass. Histologic examination of the biopsy specimen revealed Burkitt lymphoma, which was treated with chemotherapy leading to resolution of her menorrhagia.
Summary and Conclusion: Burkitt lymphoma presenting as a vaginal/cervical mass is exceedingly rare, especially in the adolescent patient. Burkitt lymphoma is generally highly responsive to chemotherapy, and symptoms rapidly improve after initiation of treatment.
Key Words: Vaginal mass, Burkitt lymphoma, Vaginal bleeding

Introduction

Menorrhagia is a common gynecologic complaint among adolescents that is often attributed to the developing hypothalamic-pituitary-ovarian axis or underlying bleeding diathesis,[1,2] and is rarely secondary to malignancy. In adolescents, vaginal and cervical masses are extremely rare, and the most common isolated histologies include rhabdomyosarcomas, germ-cell tumors, or clear-cell adenocarcinomas.[3] Primary non-Hodgkin lymphoma (NHL) can mimic gynecologic malignancies with masses in any organ of the female genital tract in adults.[4,5] Although lymphomas frequently manifest as lymphadenopathy, it is estimated that up to 35% of adult NHL patients present with a primary extranodal lymphoma, and that only 1.5% of extranodal NHLs originate in the female genital tract.[6,7]

To our knowledge, there are few case reports in the literature of Burkitt lymphoma presenting as a vaginal mass in adolescents. In this case report, we describe the case of a 15-year-old adolescent with menorrhagia and vaginal/cervical mass that was found to be Burkitt lymphoma. We review this patient's clinical course and the diagnosis and management of a gynecologic presentation of Burkitt lymphoma.

Case

An otherwise healthy 15-year-old adolescent presented with new onset heavy vaginal bleeding. The patient had previously had regular menses occurring every 4 weeks; however, for the 6 weeks before presentation she experienced heavy bleeding every 2 weeks. An oral contraceptive was started with some initial improvement in bleeding. She then returned to the hospital 2 weeks later passing large clots, at which point her oral contraceptive pills were titrated to be taken every 8 hours for 1 day and then tapered down to daily. This initially improved symptoms; however, her heavy bleeding recurred and she became anemic with a decrease in her hemoglobin from 13.7 g/dL to 8 g/dL.

Because of her decrease in hemoglobin, she was admitted to hospital, and again her oral contraceptive pill was increased to 3 times a day with improvement in her bleeding. Of note, the patient had 2 pelvic ultrasound examinations within the past 6 months before the development of any heavy bleeding for a hemorrhagic ovarian cyst that had resolved and no evidence of uterine/vaginal/cervical mass was noted on these images. Her vaginal bleeding during this admission was thought to be secondary to anovulatory cycles.

One month later, the patient presented to the emergency department with heavy bleeding in the setting of tapering off oral contraceptive pills. She was found to be tachycardic, hypotensive, and anemic with a hemoglobin of 6.8 g/dL. She was admitted to the intensive care unit for close monitoring and blood transfusion. She again stabilized with high-dose oral contraceptive pills and was discharged home. She then returned to care 3 weeks after this admission with continued heavy vaginal bleeding and a pelvic ultrasound was obtained after gynecologic consultation. The pelvic ultrasound revealed a new well circumscribed, heterogeneously hypoechoic and hyperemic mass measuring approximately 8.0 × 8.0 × 7.1 cm at the level of the cervix

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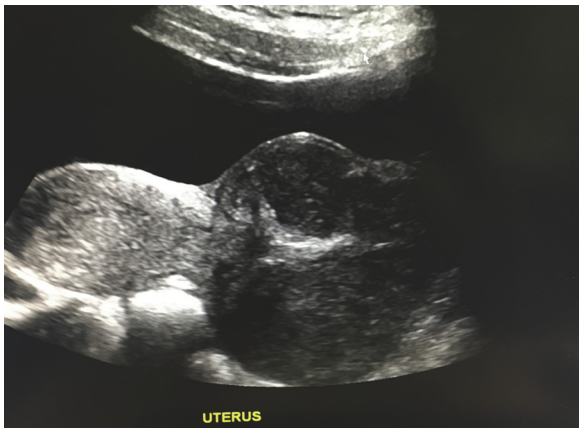

Fig. 1. Pelvic ultrasound examination was done on final evaluation before the first operative examination under anesthesia and biopsy. The image shows a new vaginal/cervical mass measuring 8 × 8 × 7.1 cm.

and protruding into the vagina, which was highly concerning for a solid tumor (Fig. 1). The patient had never had a pelvic exam during her presentations because of her age.

The patient was taken to the operating room for an exam under anesthesia and excisional biopsy of the distal portion of the vaginal mass. Rectal exam revealed a large, firm, 8-cm vaginal mass. On speculum exam, there was a large mass filling the vagina, with the apex of the mass unreachable on digital exam. A resection of the distal section of the mass was performed with the use of electrocautery to maintain hemostasis. Intraoperative frozen section was obtained, which resulted in a preliminary diagnosis of lymphoma. At the conclusion of the excisional biopsies, the tumor bed was coated with ferrous subsulfate to decrease the risk of postoperative bleeding.

Final pathologic review of the specimen revealed a Burkitt lymphoma with a classic CD20 and Ki-67 immunohistochemical positivity, CD10, CD20, and lambda-positive using flow cytometry, and c-MYC rearrangement using fluorescence in situ hybridization. Physical examination was notable for a 3-cm left breast mass that had been growing for 6 weeks, a 0.5-cm left chest wall mass, and multiple small nodules in the right breast. Staging evaluations with neck-pelvis computed tomography scans revealed a 2.4 × 2.1-cm left iliac node, 2.8 × 1.8-cm left breast mass, and multiple smaller nodules in both breasts as well as one 8.4 × 8.2-cm vaginal mass. A whole-body positron emission tomography scan showed increased fluorodeoxyglucose avidity in a large heterogeneous mass in the vagina, multiple foci in bilateral breasts, soft tissues of the anterior chest wall, bilateral iliac chain lymph nodes, and diffuse uptake in the bone. Bilateral bone marrow aspiration and biopsy showed 5%-10% marrow involvement with lymphoma. Cerebrospinal fluid was negative for disease.

A discussion of surgical resection was had with the patient and her family for bleeding control. After multidisciplinary meetings, surgical resection was not believed to be necessary because Burkitt lymphoma is usually highly response to chemotherapy. She started treatment as per the French-American-British/Lymphomes Malins B backbone therapy for treatment with the additional use of rituximab.[8–10] After 1 cycle of reduction therapy with low-dose cyclophosphamide, vincristine, and prednisone, repeat imaging on day 8 showed a decrease in the size of the vaginal mass to 5.8 × 5.4 × 5.7 cm with resolution of the necrotic-appearing iliac node and breast mass. A repeat bone marrow biopsy showed no evidence of lymphoma. Vaginal bleeding resolved by day 5 of treatment.

She then received 2 induction cycles with rituximab, cyclophosphamide, vincristine, prednisone, doxorubicin, and high-dose methotrexate and 1 consolidation cycle with rituximab, cytarabine, and high-dose methotrexate. Positron emission tomography and computed tomography scans to assess for remission were performed 10 weeks after beginning chemotherapy showed a residual cervical/vaginal mass measuring 3 × 4 cm with fluorodeoxyglucose avidity. On the basis of imaging it was difficult to discern if this mass represented necrotic residual tissue vs active lymphoma. Therefore, the patient underwent another exam under anesthesia with distal mass excision of 1.5-cm of visible tumor arising from the cervix. The final biopsy pathology showed no active lymphoma cells with only necrotic tissue and inflammatory infiltrate. The patient then received a final planned cycle of rituximab, cytarabine, and high-dose methotrexate to complete her chemotherapy. She has remained without evidence of disease in the 3 months since completion of treatment.

Summary and Conclusion

Burkitt lymphoma presenting as a vaginal/cervical mass is an exceedingly rare presentation that, to our knowledge, has not been previously described in an adolescent patient. Burkitt lymphoma is a highly aggressive B-cell neoplasm, which is more likely to have extranodal involvement than other forms of lymphoma.[11]

Diagnosis might be difficult because many providers are unfamiliar with this possible presentation of the disease. Its rapid growth also creates an acute presentation making diagnosis challenging. The rapid doubling time of Burkitt lymphoma within 48 hours to 1 week is well described in the literature,[12] and thus in retrospect this tumor was most likely not the etiology of her initial bleeding issues. She most likely had initial anovulatory bleeding followed by the development of a vaginal/cervical lymphoma. In general, the diagnosis of malignancy might be easily missed if a patient's bleeding is assumed to be secondary to anovulatory cycles without imaging. Pelvic exam is usually not done routinely in the adolescent population, because it often requires an exam under anesthesia. Therefore, pelvic ultrasound is often the first-line preferred diagnostic tool for vaginal, cervical, or uterine masses. It is also important to note that this patient's initial presentation might have been secondary to anovulatory bleeding because she had 2 normal pelvic ultrasound examinations at 3 and 6 months before diagnosis with no evidence of a vaginal/cervical mass on these images. However, reimaging should be considered in patients with heavy vaginal bleeding resulting in anemia, such as in this patient. In rare instances, this might lead to the detection of a mass as the source of menorrhagia.

Burkitt lymphoma is highly sensitive to chemotherapy with rapid reduction seen in tumor size. Although surgical resection is not indicated for treatment of advanced Burkitt lymphoma, surgical resection of the tumor was discussed in this patient's case to help control vaginal bleeding. However, surgery was deferred because of the potential risk of hemorrhage because of the known poorly defined infrastructure of a Burkitt tumor, which creates difficulty in putting traction on the tumor during a proposed surgical resection. Instruments placed on the tumor for stabilization and retraction do not hold in place. Because of the known excellent response of Burkitt lymphoma to chemotherapy, treatment was started immediately when diagnosis was confirmed. In this case, the patient's vaginal bleeding subsided quickly after induction chemotherapy and repeat imaging within 1 week showed an interval decrease in tumor size.

This case provides an opportunity to review the diagnosis and management of an unusual cause of menorrhagia and a new vaginal/cervical mass in an adolescent patient. The detection of Burkitt lymphoma as a vaginal mass in an adolescent is exceedingly rare, whereas menorrhagia is quite common. Further investigation with ultrasound examination of refractory menorrhagia thought to be secondary to anovulation should be considered in all adolescents.

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