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Appendiceal and ovarian Burkitt's lymphoma presenting as acute appendicitis

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ABSTRACT

Burkitt's lymphoma is an extremely aggressive B-cell non-Hodgkin lymphoma. Patients with the sporadic form of Burkitt's lymphoma typically present with a rapidly growing abdominal mass, pain and distension. Involvement of either the appendix and/or ovaries in females is a rare manifestation of the disease. We present an unusual case of a 13 year old girl with appendiceal and ovarian Burkitt's lymphoma presenting with signs of acute appendicitis. This case demonstrates the potential for secondary involvement of the appendix and/or ovaries from Burkitt's lymphoma as well as the importance of the histopathology.

1. Introduction

Childhood cancers consist primarily of leukemias, lymphomas, sarcomas, and cancers of the central nervous system. Lymphomas (both Hodgkin and non-Hodgkin) are one of the most common pediatric malignancies in the United States, encompassing 16% of childhood cancers. Non-Hodgkin Lymphomas, a subset of all lymphomas, comprise approximately 5% of all childhood cancers [1–3]. Burkitt's lymphoma is an extremely aggressive form of mature B-cell non-Hodgkin lymphoma. It was first described by surgeon Denis Burkitt in Kampala, Uganda as a sarcoma of the jaw. Later, it was reclassified as an aggressive form of lymphoma and contributed greatly to the study of Ebstein Barr virus (EBV) for which it was commonly associated with [4,5,7]. There are now three clinical forms of Burkitt's Lymphoma: endemic, sporadic, and immunodeficiency-associated [6].

Patients presenting with Burkitt lymphoma have symptoms depending on the type and location. Patients with the endemic form have involvement of the facial bones and jaw. The sporadic form often presents with rapidly growing abdominal masses. Finally, the immunodeficiency associated form is typically associated with diffuse lymphadenopathy. Extranodal sites of involvement include the kidneys, gastrointestinal tract, omentum, ovaries, and breast [2,4]. We present an unusual case of a pediatric female with appendiceal and ovarian Burkitt lymphoma presenting with signs of appendicitis.

2. Case report

The patient is a 13 year old girl that presented to the emergency department with acute right lower quadrant abdominal pain. The patient was well until the day before when she developed abdominal pain and subjective fever. The pain was constant, non-radiating, and worsened with walking. The pain was associated with nausea and non-bilious emesis without diarrhea or constipation.

On presentation, she was afebrile with mild tachypnea; all other vital signs were within normal limits. Physical exam revealed an alert patient with a soft and non-distended abdomen with diffuse tenderness to palpation, particularly localized to right lower quadrant. Laboratory results revealed a leukocytosis to 15.1 × 103/mcL with 93% neutrophils and elevated C-Reactive Protein to 2.0 mg/dL. All other laboratory values were within normal limits.

Abdominal and pelvic ultrasound showed findings consistent with acute appendicitis and a non-visualized left ovary with a complex left adnexal mass measuring 8.0 × 6.0 × 7.0 cm and small amount of free fluid within the pelvis. A CT scan was subsequently performed which demonstrated a 6.8 × 2.4 × 2.7 cm tubular, dilated, fluid-filled, blind ending structure arising from the cecum with surrounding inflammatory changes indicative of appendicitis as well as a solid and cystic indeterminate left ovarian mass (Fig. 1A and B).

Given the CT findings, tumor markers were sent and the patient was initially treated with IV antibiotics. Normal AFP and quantitative βhCG levels were obtained, and the patient was taken to the operating room for a laparoscopic appendectomy and ovarian sparing surgery with

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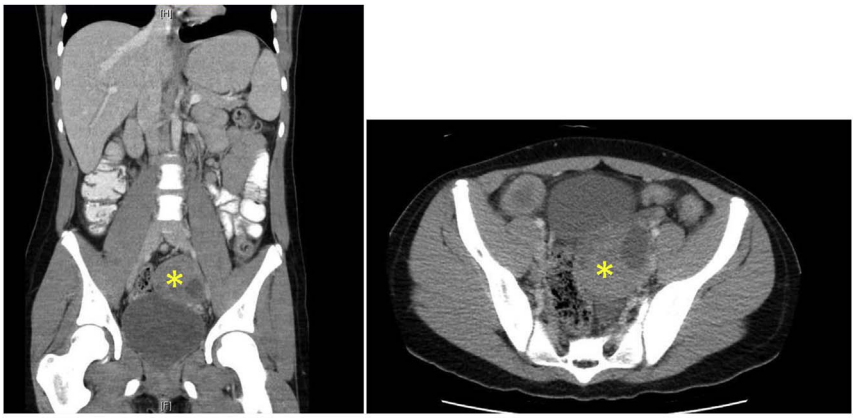


(A) (B)

Fig. 1. A – Representative Coronal CT scan image demonstrating 8.0 × 6.0 × 7.0 cm left ovarian mass (Asterix on ovarian mass). B – Representative Axial CT scan image demonstrating an enlarged appendix and the solid and cystic left ovarian mass (Asterix on ovarian mass).

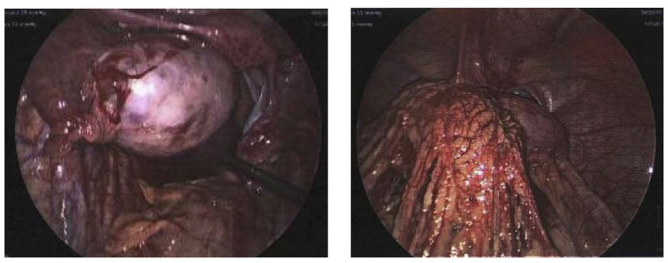
possible oophorectomy.

Operative findings revealed a distended, thickened appendix with adhesions to the omentum and an enlarged left ovary with no cyst wall capsule (Fig. 2A and B).

Due to the normal ovarian tumor markers and finding of what was thought to be fat on the ultrasound of the ovarian mass, the initial suspicion was for a dermoid cyst that could be resected in an ovarian sparing fashion. However, since no cyst was identified, an intra-operative biopsy of the ovary was submitted to pathology for frozen section. This revealed a small round blue cell tumor. Thus, a left salpingo-oophorectomy was performed to obtain adequate tissue for pathology and to prevent any spillage of tumor following the family's consent.

The macroscopic pathology exhibited an enlarged, dilated appendix measuring 13.8 cm in length and ranging in diameter from 0.8 cm at the appendiceal base to 3.7 cm at the appendiceal tip. The dilated portion was completely replaced by a pale pink and glistening fleshy cut surface. Evaluation of the left fallopian tube and ovary revealed the fimbriated end of the tube to be unremarkable with an adjacent 0.6 cm paratubal cyst. The enlarged ovary measured 6.9 × 5.8 × 4.7 cm and was almost entirely replaced by a pale pink-gray fleshy cut surface with focal areas of congestion (Fig. 3A and B).

Microscopic examination of the specimens supported a diagnosis of Burkitt lymphoma involving the entire appendiceal circumference and ovary as well as the fallopian tube. Tumor cells were positive for CD20 (a B-cell marker which is positive for the neoplastic cells), CD10, BCL-2 (is typically negative in Burkitts lymphoma but positive in this case indicating the presence of an additional BCL2 breakpoint consistent with a high-grade B-cell lymphoma), c-Myc (indicates a high grade B cell lymphoma), BCL-6 (germinal center marker positive in Burkitt lymphoma) and PAX-5 with a Ki-67 proliferation index of virtually 100% (indicating a high proliferation index which is typical for Burkitt lymphoma) (Fig. 4). Karyotyping from the ovarian mass was normal for t(8; 14).



(A) (B)

Fig. 2. A – Intra-operative picture of Left ovarian mass. B – Intra-operative picture of enlarged and dilated appendix with adhesed omentum.

After the pathology was finalized, the patient underwent port placement with bone marrow aspirate and biopsy and lumbar puncture. Bone marrow aspirates and core biopsies were negative for malignancy. A subsequent PET/CT of the abdomen revealed retroperitoneal lymphadenopathy at the periaortic, iliac, and pelvic regions as well as a right side pelvic mass along the pelvic wall. Staging for her B-cell lymphoma was at low risk group B which is equivalent to stage III. The patient is being treated with chemotherapy, using Children's Oncology Group Study - ANHL1131 standard arm comprised of the COPADM regimen (Cyclophosphamide, Oncovin, Prednisolone, Adriamycin, and Methotrexate without Rituximab).

3. Discussion

Children presenting with lymphomas typically have extranodal disease. Burkitt lymphoma in these patients typically present with rapidly growing abdominal masses and often with signs of an associated obstruction. Involvement of either the ovaries or appendix is a rare manifestation [9]. It is more likely that appendiceal and ovarian sites are areas of secondary involvement; although, it has been suggested that there is some pre-existing lymphoid tissue in the ovary that creates an at-risk site for lymphoma development [10]. Initial presentation of Burkitt lymphoma as appendicitis in children is an uncommon occurrence, but is a possibility since Burkitt lymphoma can affect the ileocecal region [8,14]. Appendiceal Burkitt lymphoma displays an enlargement of the appendix on CT, but without any inflammatory or infectious features. In addition, non-specific appendiceal enlargement due to Burkitt lymphoma could play a role in the pathogenesis for acute appendicitis. Pain in the right lower quadrant mimicking acute appendicitis can also be from an ileocecal intussusception due to Burkitt lymphoma [8,27].

Solid ovarian tumors need to be distinguished from lymphomas via histologic and biologic markers such as cytokeratin, lactate dehydrogenase, alpha fetal protein and human chorionic gonadotrophin. Identification of germ cell tumors alters the management and chemotherapeutic treatment [24,25]. A key suspicion for the development of an ovarian Burkitt lymphoma is symptoms of a rapidly growing mass in the pelvis. Primary ovarian lymphomas account for 0.5% of lymphomas and are involved in 19% of all adnexal lymphomas. They are defined by Fox and Langley as disease confined to the ovary, absence of disease in blood and bone marrow, and possibility for extraovarian deposits only after a few months. The prognosis for Burkitt lymphoma with ovarian involvement can be worse, with higher rates of death due to the disease within the first year of diagnosis [11,24].

A diagnosis of Burkitt lymphoma is made from a surgical biopsy based on immunohistochemical patterns from tissue and marrow biopsies. Biopsies of the tumor mass with fine needle aspiration (FNA) can provide rapid diagnostics, but typically additional surgical biopsy