A 12-Year-Old Girl with Persistent Abdominal Pain and Emesis

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A previously healthy 12-year-old girl presented to the emergency department in May 2013 with diffuse abdominal pain and vomiting. She was evaluated with ultrasounds of the ovaries and appendix, both of which were negative except for mesenteric adenitis. An X-ray of the abdomen was normal. She was discharged with a diagnosis of mesenteric adenitis. Her symptoms resolved within 2 days and she left for a month-long family vacation to Malaysia, Laos, Singapore, and Indonesia. On June 15th, while on vacation, she experienced abdominal pain without vomiting for 2 days, which resolved. She returned to the United States on June 26th, and the abdominal pain recurred on July 1st.

On July 3rd she was admitted to the emergency department with intractable bilious vomiting and diffuse abdominal pain. Her abdominal pain was rated as 8 of 10 and was described as waxing, waning, sharp, stabbing, and localized to the right lower quadrant. She denied loss of appetite, changes in bowel habits, or chest pain. She indicated that the pain was exacerbated by movements and walking. Her parents reported that the antiemetic medications did not relieve the pain, and that due to her vomiting she had minimal food intake. Her past medical history was significant only for mesenteric adenitis in May 2013, with no past surgical procedures. A review of symptoms was negative for recent illness, constipation, diarrhea, palpitations, shortness of breath, or menstrual complications. All of her immunizations were up to date, she had no known food or drug allergies, and was currently being administered ondansetron. Her family history was only significant for kidney stones. The patient denied a history of Crohn’s disease, aortic aneurysm, gall stones, or ulcerative colitis. Social history was noncontributory.

On physical examination in the emergency department, the patient was moaning and ill-appearing. Vital signs were temperature of 37.4°C, heart rate of 89 beats per minute, respiratory rate of 19 breaths per minute, blood pressure of 101/63 mm Hg, and oxygen saturation of 100% on room air. The patient had dry mucosal membranes, pallor, and abdominal tenderness along the right lower quadrant. There were normal active bowel sounds and no appreciable organomegaly. Upon deep palpation, a mass was found in the right lower quadrant near the periumbilical region that was firm, nonmotile, and nonpulsatile. No other significant physical examination findings were noted. Upon completion of the physical examination, the patient had another episode of green, bilious emesis.

Initial complete blood count with differential study abnormalities included white blood cell count of $7.76 \times 10^3$ cells/L with an elevated segmented neutrophils percentage of 81%, without presence of left shift. The complete metabolic panel abnormalities were blood glucose of 69 mg/dL and a CO$_2$ level of 19 mmol/L. There was an elevated erythrocyte sedimentation rate of 24 mm/h. The urinalysis revealed elevated albumin of 1+, and ketones of 3+, otherwise normal. The remainder of her complete blood count, complete metabolic panel, acute phase proteins, and urinalysis results were all within normal limits.

The patient had a computed tomography (CT) scan of the abdomen and pelvis that indicated the presence of an intussusception (Figure 1).

She was given two normal saline intravenous fluid boluses for dehydration. Ondansetron was administered for emesis, but was ineffective. A switch to promethazine was made for emesis control. A pediatric surgeon was consulted for exploration and open reduction of the intussusception. The intussusception was reduced but a large cecal mass was present without mesenteric adenopathy or obvious intra-abdominal pathology; it was sent to pathology for review.

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Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.
Diagnosis:
Burkitt’s Lymphoma of the Ileocecal Junction with Uninvolved Mesenteric Lymph Nodes

The patient was diagnosed as having Burkitt’s lymphoma (BL) as the pathologic lead point for the intussusception. The immunohistochemical profile of the mass was CD10, CD19, CD20, and PAX5 positive. She was referred to pediatric oncology after the diagnosis and a metastatic work-up was completed, including positron emission tomography, CT of the pelvis and chest, bone marrow studies, and cerebrospinal fluid (CSF) analysis, all of which were negative. The mass was diagnosed as a stage I tumor. The patient was treated with two courses of methotrexate, cytoxan, doxorubicin, vincristine, prednisone, intrathecal methotrexate, and hydrocortisone. Two courses of rituximab (anti-CD20) were also included because the tumor was CD20 positive. The importance of intrathecal chemotherapy was emphasized to reduce the risk of CSF disease. The patient had adverse effects of eye irritation, redness, and mild upper eyelid swelling followed by bilateral styes after the first cycle of chemotherapy. She recovered well without major complications during treatment.

DISCUSSION
In healthy children, BL has an annual incidence of less than 300 cases in the United States. It is an aggressive, mature B-cell lymphoma that occurs in patients of all ages and characteristically has one of the shortest doubling times of all human cancers.1

There are three distinct epidemiologic clusters of BL. Endemic BL is commonly found in children living in sub-Saharan Africa and New Guinea, is consistently Epstein-Barr virus (EBV) positive, and primarily presents in the jaw or facial bones. Sporadic BL (the diagnosis in our patient) commonly accounts for 40% of pediatric lymphoma cases, is usually found in North America and Europe, and only has a 15% association with EBV.2 This subtype has up to a 90% incidence in the gastrointestinal tract (particularly the ileocecal region), with other cases developing in the testes, ovaries, nasal sinuses, and lymph nodes.3 Immunodeficiency-associated BL (IABL) is often associated with HIV infection and is commonly one of the initial diseases associated with the manifestation of AIDS.4 IABL may occur in posttransplant patients taking immunosuppressive drugs.

On microscopy, this tumor usually consists of lymphoid cells with a high mitotic rate and that are said to have “starry sky” appearance due to scattered macrophages containing dead tumor cells.5 Immunohistochemical and genetic analysis are essential for the diagnosis and treatment of BL. The tumor cells generally express markers of B-cell differentiation, as seen in our patient who was CD10, CD19, and CD20 positive, which is a therapeutic indication for the use of rituximab.6 On genetic analysis, all types of BL show dysregulation of the c-myc gene by one of three translocations, the most common being t(8;14)(q24;32), which occurs in about 80% of cases.7

Management of this malignancy is vital, as the rapid doubling rate of this cancer can cause an oncologic emergency. Staging commonly involves multiple imaging modalities, such as chest radiography, CT of the chest, abdomen, bone marrow biopsy, and CSF analysis, to evaluate for metastatic disease. In this patient, all of the aforementioned tests were negative. Blood tests should be performed to assess for any urgent electrolyte abnormalities, along with concurrent HIV and EBV tests.

BL has a high event-free survival rate in children (~90%)8 with two or three relatively short courses of multiagent chemotherapy (as was administered to our patient).1 Intrathecal therapy is an essential component of
treatment for all patients to reduce likelihood of CSF invasion. Our patient had stage I BL with negative margins upon resection of the cecal mass. She had minimal adverse effects during therapy and has no residual long-term side effects 7 months posttherapy.

CONCLUSION

Intussusception in an adolescent is rare, and its presence requires broadening the differential diagnosis to include the possibility of a malignancy as the lead point. Sporadic BL is an aggressive cancer that should be considered as a diagnosis in a patient with a history of recurrent intractable vomiting and abdominal pain, and it may be palpated with a thorough physical examination. Our case represents a unique characterization of this rare malignancy in the pediatric population. It emphasizes the need for thoroughness when obtaining a review of systems while maintaining a broad differential with the presence of prolonged and inexplicable emesis.

REFERENCES