ACTA SCIENTIFIC GASTROINTESTINAL DISORDERS

Volume 2 Issue 3 May 2019

Clinical Image

An Unusual Cause of Upper Gastrointestinal Bleeding in a Young Patient

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Received: March 28, 2019; **Published:** April 18, 2019

A 20-year-old man who had recently sustained a sacral fracture while lifting a television presented with epigastric pain and melena. His course began one month prior when he developed back pain, and he was started on nonsteroidal anti-inflammatory drugs (NSAIDs) and steroids. As his back pain persisted, he underwent an MRI of the spine revealing a sacral fracture and a density in the right piriformis concerning for a hematoma versus a mass. Notable initial labs revealed hemoglobin 6.8, white blood cells 14, platelets 502, albumin 1.6, total protein 3.2, and INR 1.1. He underwent EGD for evaluation of the epigastric pain and melena that revealed numerous one- to two-centimeter polypoid masses with heaped-up edges and central ulcerations throughout the stomach (Figure 1 and Figure 2). Pathology from biopsies of these gastric lesions as well as from a CT-guided biopsy of the piriformis mass confirmed Burkitt's lymphoma. A CT scan confirmed stage IV disease and revealed an entero-enteric intussusception (Figure 3). HIV testing was negative. He was subsequently started on chemotherapy with hyper-CVAD and intrathecal cytarabine and is in remission over one year after completion of therapy. He did not have any further evidence of overt gastrointestinal bleeding and his epigastric pain resolved, with subsequent cross-sectional imaging not showing evidence of intussusception, suggesting that it resolved spontaneously. To date, he has not required surgical intervention.

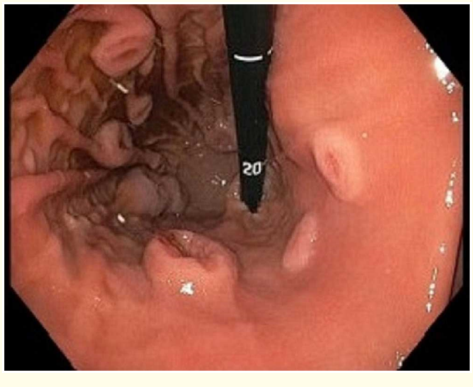

Figure 1: EGD showing gastric polypoid masses throughout the stomach on retroflexion.

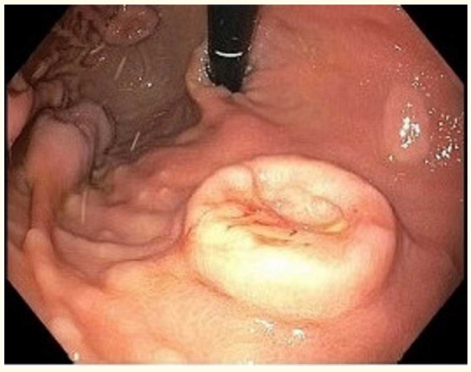

Figure 2: EGD showing Burkitt's lymphoma of the stomach.

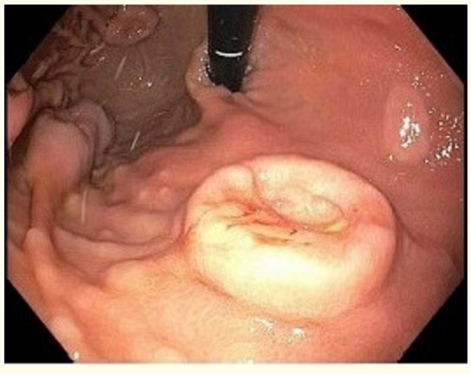

Figure 3: CT scan of the abdomen demonstrating a small bowel intussusception.

Burkitt's lymphoma is a very rare cancer, comprising only 1-2% of lymphomas, though its incidence is rising [1,2]. Although extranodal involvement in Burkitt's lymphoma is rare, the gastrointestinal tract is the most common site where it occurs [3], leading to a myriad of gastrointestinal symptoms that are important for clinicians to recognize including abdominal pain, obstruction, intussusception, gastrointestinal bleeding, and a palpable mass [1,4]. Systemic symptoms can include, as with other types of lymphoma, night sweats, fevers, and weight

loss, as well as effects from involvement of the spleen, liver, ovaries, breasts, bone marrow, and central nervous system [4]. The sporadic variant of Burkitt's lymphoma seen in the United States and Western Europe most commonly presents as an intra-abdominal mass [5] and has an incidence of 2.2 cases per million per year [6]. It occurs more commonly in children, accounting for approximately 40% of pediatric lymphomas [7]. The endemic form of Burkitt's lymphoma, most common in Africa, typically presents as a jaw tumor. It has a much higher incidence around 3-6 per 100,000 children per year and is strongly associated with EBV infection.[5] Finally, the immunodeficiency-associated variant is most commonly seen in patients with HIV, and there is a 261-fold increased risk of Burkitt's lymphoma in this population [8]. It comprises 30-40% of all non-Hodgkin's lymphoma in HIV-positive patients, though it can also occur in post-transplant patients and patients with immunodeficiency [7]. When gastrointestinal involvement of Burkitt's lymphoma is present, the stomach is involved most frequently, comprising 50-60% of gastrointestinal extranodal sites, followed by the small and large intestines, encompassing 30% and 10% respectively [1,2]. Few cases have been described of patients with Burkitt's lymphoma presenting with gastrointestinal tract involvement as evidenced by concurrent melena and intussusception as occurred in this case. Additionally, many cases described have occurred in HIV positive patients [2-4,9]. It is important to consider this diagnosis in young patients presenting with abdominal pain and melena, especially in the setting of a fragility fracture, as these findings are unusual in young patients.

The differential diagnosis of gastric lesions that may appear similar to the endoscopic findings seen in our patient include diffuse large B-cell lymphoma, MALT lymphoma, T-cell lymphoma, Mantle-cell lymphoma, and metastatic disease [10]. The optimal treatment of Burkitt's lymphoma has not been determined due to few randomized studies but generally consists of multiagent chemotherapy with central nervous system prophylaxis. Additional treatments include rituximab, steroids, radiation, and stem cell transplantation.[11] Surgery is not the mainstay of therapy but is reserved for complications such as refractory obstruction, bleeding, or perforation due to lymphoma. Regarding prognosis, prompt treatment usually cures the disease in children, with long-term survival rates ranging from 60% to 90%. In adults, rapid initiation of treatment leads to survival rates around 70-80% [7].

In conclusion, it is important for clinicians to maintain a broad differential diagnosis in such cases, as management and urgency of diagnosis differs significantly from more common pathologies such as peptic ulcer disease. Additionally, it is vital that clinicians recognize the diverse signs with which Burkitt's lymphoma can present, as the incidence is increasing, and it is important to make the diagnosis and initiate treatment promptly so as to ensure the best possible prognosis.

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