Plasmablastic Lymphoma Causing Adult Intussusception After Cardiac Transplantation

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ABSTRACT

Intussusception in adults is a rare occurrence at approximately 5% and malignancy as the cause comprises half that number. The most common malignancies found are primary adenocarcinoma, metastatic carcinoma, lymphoma, and gastrointestinal stromal tumors. Lymphoma is the second most common. The management of adult intussusception is generally surgical, which is due to the higher likelihood of malignancy being the underlying cause. The patient's history helps to direct management and the most likely underlying diagnosis. This is especially important in patients who are immunosuppressed and with a history of lymphoproliferative disease. Early management and proper surgical intervention allow for the best survival rate. Here we present a case of adult intussusception caused by a rare and aggressive type of non-Hodgkin lymphoma.

Key Words: Adult, Intussusception, Lymphoma, Plasmablastic.

INTRODUCTION

Intussusception, the invagination of a bowel segment into an adjacent one, is an exceedingly rare phenomenon in adults.[1] In adults the likelihood of a malignant process causing intussusception is high.[2] From the various malignancies, lymphoma being the causative pathology is rare and for the most part discussed in isolated case reports. It is important to realize the potential severity of the underlying malignant process. We believe it is important to continue reporting these rare occurrences to increase our understanding of the disease process, presentation, and treatment options. Here we present a case of plasmablastic lymphoma, an aggressive non-Hodgkin lymphoma, causing adult intussusception after cardiac transplantation.

CASE REPORT

A 26-year-old White female was admitted with a three-day history of diffuse, colicky abdominal pain that was most severe in the right lower quadrant. She also endorsed nausea and new onset hematochezia.

The patient's medical history was significant for restrictive cardiomyopathy, for which she underwent an orthotopic heart transplant at age 11. She developed stage II Hodgkin like post-transplant lymphoproliferative disease several years following transplantation in the setting of chronic immunosuppressive therapy. She had completed three cycles of chemotherapy and had been, at the time of current presentation, in remission for five years. Her current immunosuppressive regimen included cyclosporine and sirolimus.

She was hemodynamically stable at the time of presentation. Physical examination revealed a thin, nontoxic appearing young woman. Her abdomen was soft, nondistended, and tender in the right lower quadrant with localized guarding. She had no other significant physical examination findings. Initial laboratory results showed a normal white blood cell count. A basic metabolic panel revealed a creatinine of 1.4, however she has a known history of renal insufficiency with a baseline creatinine of 1.3.

Computed tomography scan demonstrated ileocolic intussusception with associated mesenteric edema and free fluid, concerning for possible ischemia with no lymphadenopathy (**Figure 1**). Subsequently, the patient was brought to the operating room for diagnostic laparoscopy. The terminal ileum was traced from distal to proximal starting at the cecum; intussusception in the terminal

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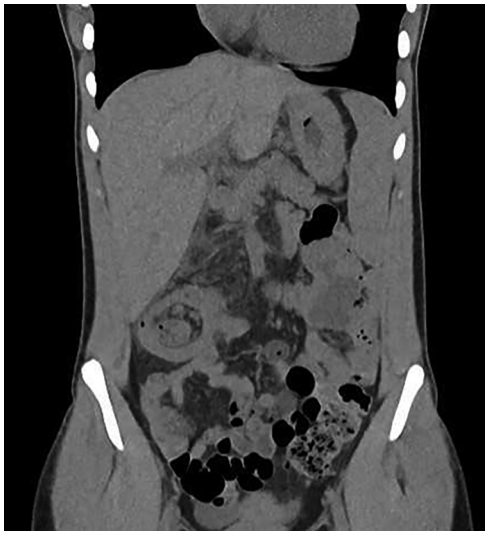

Figure 1. Computed tomography abdomen and pelvis, coronal section, showing target sign.

ileum was confirmed (**Figure 2**). Additionally, an enlarged, associated mesenteric lymph node was identified. The patient then underwent a laparoscopic right hemicolectomy and mesenteric lymph node excision. Dissection began with identification ileocolic pedicle and high ligation with an Endo GIA stapler. This was followed by a lateral to medial mobilization of the right colon. As the patient was quite thin, and of relatively short stature, a Pfanenstiel incision was chosen as the extraction site, after sufficient mobilization, through which the specimens were delivered with ease. Examination of the resected bowel revealed a 3.9 cm mass within the cecum (**Figure 3**).

The patient was discharged to home on postoperative day three in stable condition, having suffered no postoperative complications. She was seen in the outpatient setting two weeks later at which point she continued to do well. Immunohistochemical analysis were positive for CD138, CD45, and CD10 and negative for CD20. The Ki-67 proliferation rate was 80% – 90%. The anaplastic large cell lymphoma kinase and human herpes virus were both negative. She was diagnosed with plasmablastic lymphoma.

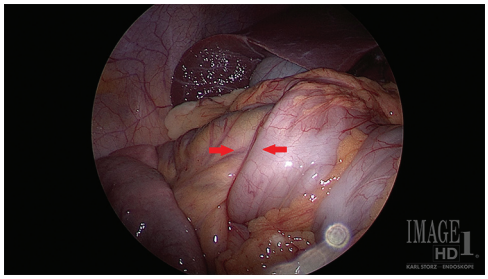

Figure 2. Intraoperative view of the intussuscept bowel.

DISCUSSION

Intussusception is an invagination of a segment of bowel into the lumen of an adjacent segment. It is rare among adults; in fact, only 5% of all cases are found within this population.[1] Reportedly, at least 50% of these are secondary to an underlying malignancy.[2] In the current surgical literature, there are only 36 reported cases within an 11-year period that attribute lymphoma as the causative etiology in adult ileocolic intussusceptions[4] – elucidating the rare nature of this pathologic entity. The most common malignancies found are primary adenocarcinoma, metastatic carcinoma, lymphoma, and gastrointestinal stroma tumors, with lymphoma being the second most common.[3]

Currently, the mainstay of management for adult intussusception is surgical. This is in part due to the higher likelihood of malignancy as being the underlying cause. Notably, there have been reports of nonoperative management for adult intussusception, but one must take into consideration the patient's clinical circumstance.[10] Patient history indicating a higher likelihood of malignancy and clinical findings concerning for obstruction or ischemia should direct surgical management. In our case, a history of post-transplant lymphoproliferative disorder as well as a classic presentation for intussusception, in addition to convincing radiologic evidence of this disease process, heightened our clinical suspicion for an underlying malignant process. For ileocolic intussusception, it is important to consider whether to attempt reduction or not. Signs or concern for ischemia and or gangrene should preclude one from attempting reduction. Colonoscopy can be helpful

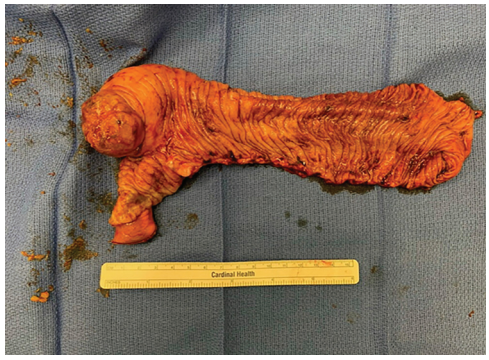

Figure 3. Back table view of the gross specimen showing the mass.

in determining if the lesion at the lead point is benign or malignant,[11] but should not supplant or delay necessary surgical therapy (which usually entails resection). Treatment for lesions highly suspected to be benign may begin with attempts at reduction and local excision. Lesions that are suspicious for malignancy or segments that cannot be reduced should be resected en bloc with the supplying mesentery. Our case illustrates that a minimally invasive approach is safe and feasible in such scenarios. Furthermore, a Pfanenstiel extraction site (when anatomically permissible) confers a more cosmetic scar in young patients and is associated with lower rates of incisional hernia, particularly in patients with compromised wound healing secondary to chronic immunosuppressive therapy in the setting of organ transplantation.

Our patient's pathology revealed a rare, highly aggressive, non-Hodgkin's lymphoma known as Plasmablastic lymphoma (PBL). PBL was first described in the oral cavity/jaw of HIV infected patients. However, there are increasing cases of HIV-negative PBL found in extraoral sites including the gastrointestinal tract.[5] PBL is male predominant, most frequently occurring in the fourth decade of life in HIV positive patients, compared to HIV negative patients, in which is occurs more commonly during the sixth decade.[6]

PBL is associated with other causes of immunosuppression such as post-transplant state, as seen in our patient.[6] Chronic immunosuppressive therapy, Epstein Barr virus (EBV) and genetic susceptibility may contribute to the increased risk of developing PBL[6] Our patient's history included both sirolimus and cyclosporine therapy; each drug increases the risk of lymphoproliferative disease[7,8] by 1% to 6%.

PBL is an aggressive tumor with poor prognosis. The heterogenous manifestation of this lymphoma renders its treatment challenging. Presently there is no standard of care; multiple chemotherapy regimens such as cyclophosphamide, doxorubicin, vincristine, and prednisone have been associated with different outcomes. Patients with complete clinical response have a median overall survival of 48 months.[6] However, without complete response the median overall survival was found to be three months.[6] Other modalities include autologous stem cell transplant, immunomodulators, and antiviral agents and EBV-targeted therapy, but further studies are needed in these areas.[9]

We report a case of Plasmablastic lymphoma as a cause for intussusception in an immunocompromised, postcardiac transplant individual who is HIV-negative. As this is a highly aggressive tumor with no current standard of care, it is important to recognize a patient's clinical history in order to assist in diagnosis. The high incidence of intussusception in adult malignancies should be kept in mind. In order to achieve the best survivability, early surgical management and oncologic treatment are required for this rare and aggressive disease.

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