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Myeloid Sarcoma: A Rare Cause of Small Bowel Obstruction

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ABSTRACT

Myeloid sarcoma is a rare and aggressive tumor of myelogenous origin that uncommonly presents in the small bowel. Here we describe the case of a 29 year old male with a two week history of worsening abdominal pain associated with nausea and bilious vomiting. The patient displayed tenderness in the epigastric and periumbilical regions with grossly normal lab values. On CT he was found to have a small bowel obstruction with a transition point in the pelvis. Exploration revealed a small bowel mass located approximately 12 cm from the ileocecal valve. Pathological analysis showed large malignant cells with a high mitotic rate that were positive for CD 45, CD43, CD34, CD117, and BCL2. Also in patients that ultimately recover, the length of complete remission is also very low. Unfortunately, due to its rarity, MS can often be misdiagnosed as a more common gastrointestinal tumor until pathology is finalised. Additionally signs and symptoms for this tumor can be vague, unless causing bowel obstruction. In this case study we present a 29 year old male with obstructive symptoms whom was found to have a pelvic mass that was ultimately diagnosed as MS.

INTRODUCTION

Myeloid Sarcoma (MS) is a very rare and aggressive tumor originating from cells of myelogenous origin [1]. It is also known as Chloroma due to its green color of appearance from the extensive expression of the myeloperoxidase enzyme (MPO) [1]. The tumor is usually comprised of cells that develop from myeloid cell lineage and range from different levels of maturity from immature, in which the cell are undifferentiated, to mature, in which the cell are more well differentiated [1]. Populations of cells within myeloid sarcoma can vary as well from eosinophils, granulocytes, myeloblast, and promyelocytes [1]. MS is more commonly found to present in the skin and lymph nodes. But has been observed in specific rare cases to occur in the oral mucosa, nasal mucosa, breast, pleura, retroperitoneum, gastrointestinal tract, and testis [1]. More specifically, only 0.6% of MS occurs in the gastrointestinal tract making its presentation in this area of the body very rare [6].

CASE PRESENTATION

29 y/o male presented to the emergency room with 2 week hx of abdominal pain, nausea, and bilious vomiting. He had previous went to the hospital 1 wk prior with similar complaints but was discharged. Pt reported constipation and decreased bowel movement and stated his last bowel movement was yesterday the day before surgery. Physical exam showed mild to moderate tenderness in the periumbilical and epigastric regions. Lab values were as follows: Lactate 1.01, WBC 8.1K, Hemoglobin 17 gm/dl, and Creatinine 1.01.

CT of the abdomen and pelvis showed small bowel obstruction with a transition point in the pelvis, intimal thickening, and mesenteric lymphadenopathy. The patient was taken to the operating room for emergent diagnostic laparoscopy. During the procedure a pelvic mass was detected 12 cm from the ileocecal junction. The operation was converted to exploratory laparotomy.

No other masses were visualized after running the length of the small bowel from the ligament of Treitz to the ileocecal junction. The segmental bowel resection of the mass with primary anastomosis was performed. Biopsies of mesenteric lymph nodes and the mass were sent for pathological analysis.

The patient was discharged home on post op day four.

DISCUSSION

• Immunohistochemistry is one of the most important pathological test to diagnose Myeloid Sarcoma.
• The most important diagnostic markers in diagnosing MS are myeloperoxidase (MPO) and CD117.
• Other markers that are commonly found expressed in MS tumors are CD99, CD68/KP1, CD68/PG-M1, lysozyme, CD34, terminal deoxy transferase (TdT), CD56, CD61, CD30, glycoporphin, CD4, CD13, and CD33.
• Studies have shown that the best treatment for MS is Systemic chemotherapy with localized surgical tumor resection and radiotherapy.
• However, the prognosis of patients that are diagnosed with MS still remains very low.
• Also in patients that ultimately recover, the length of complete remission is also very low.
• Hematopoietic Stem Cell Transplantation has been shown promising as a future treatment option for patients with MS but much more research needs to be conducted.

CONCLUSIONS

• Myeloid Sarcoma of the gastrointestinal tract only occurs in 6.5% of cases of MS.
• From our case and a review of literature MS often goes misdiagnosed due to its rarity.
• It is important to keep MS on the differential diagnosis as a rare cause of small bowel obstruction.
• Clinical index of suspicion is crucial in identifying Myeloid Sarcoma of the small bowel.

REFERENCES