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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 region 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, the tumor cells were shown to have KI67 positivity of >80%. The pathological analysis strongly supported the diagnosis of myeloid sarcoma. Given the patient's age, site of occurrence, and overall lack of previous studies on myeloid sarcoma this case is highly notable and recommended for review.

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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 6.5% of MS occurs in the gastrointestinal tract making its presentation in this area of the body very rare [6]. Unfortunately, due to its rare nature, 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 an 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.

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
- 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.

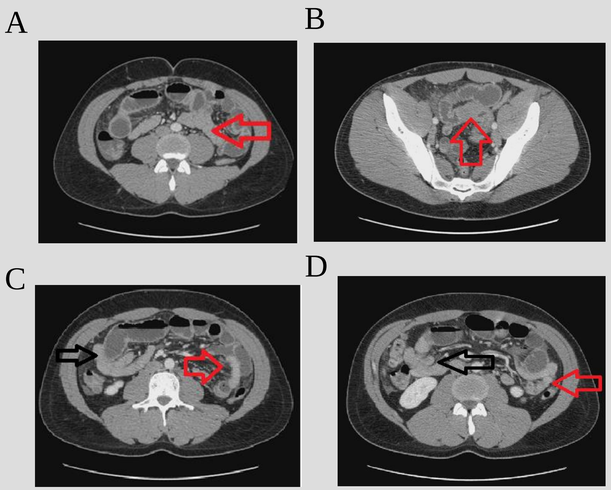

Figure 1. A) Abdominal CT scan with oral contrast showing a mass obstructing the lumen of the distal ileum with evidence wall thickening. B) Pelvic CT scan with oral contrast showing a mass obstructing the sigmoid colon. Also evidence of wall thickening and edema. C) CT scan of abdomen with oral contrast, showing multiple lymph nodes in the right and left central mesentery. Also mesenteric stranding and small bowel mass. D) CT scan of abdomen showing obstruction of terminal ileum and distal ileum.

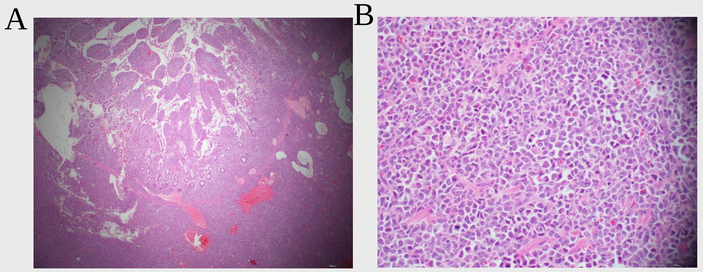

Figure 2. A) 4X magnification showing small bowel tumor. Although not shown the tumors immunohistochemical profile was positive for myeloperoxidase (MPO), CD45, CD43, CD34 CD117, and BCL2. KI67 was >80%. All strongly indicating the diagnosis of Myeloid Sarcoma. B) 40X magnification showing large malignant cells with high mitotic rate with prominent nucleoli.

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, glycophorin, 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 showing promise 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 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.

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