Case Presentation

A 61 year old male with no medical history presented to the emergency room on 3 occasions with complaints of abdominal distention, pain, nausea, and vomiting. The patient also noticed a 40 pound weight loss over last 3 months. On each occurrence he was diagnosed with a small bowel obstruction (SBO) and treated conservatively. Physical examination revealed a tender and distended abdomen. On the third occurrence, the patient presented with similar complaints and physical findings. The CBC showed a white blood cell count was 6,000 without any immature cells, hemoglobin was 14.1g/dl, and platelets were 220,000. The CMP showed a serum creatinine of 0.7mg/dl, and normal AST and ALT. The CT of the abdomen showed distended loops of bowel with air levels, adenopathy in left lower quadrant. There was also stenosis seen in the left lower quadrant. An exploratory laparotomy was performed with resection at the junction of the jejunum and ileum. Pathologic examination revealed a white fibrous tumor extending into the small bowel lumen. Initial microscopic evaluation indicated a large cell lymphoma due to cellularity. However, immunohistochimical (IHC) staining was consistent with granulocytic sarcoma (GS). Peripheral blood smear revealed no abnormal blasts and bone marrow biopsy showed no evidence of marrow involvement all indicating a primary GS (PGS). The patient was given induction chemotherapy for AML. The patient tolerated the treatment well and went into total remission.

Granulocytic sarcoma, also known as Chloroma, is a rare extramedullary (EM) manifestation of Acute Myeloid Leukemia (AML) that may occur concurrently with AML or as a sign of relapse after successful treatment. GS may present as an isolated EM tumor in an otherwise non-leukemic (NL) patient. GS is a unique form of AML. GS is an extramedullary manifestation of myeloid tumor occurring in 2-7% of patients with AML. PGS in NL patients is a rare finding with 90% progressing to AML within 10.5-11 months. Generally, GS has a predilection for the skin, bone and lymph nodes with rare cases presenting in the small intestine (~7%). The clinical symptoms of GS depend on the site of origin. In this case, the main complaints were abdominal pain and multiple SBOs. When patients present with signs of leukemic process such as thrombocytopenia, anemia, weight loss, and initial workup is negative, clinicians should consider GS on their differential diagnosis. Definitive diagnosis is made by biopsy and IHC staining. Making the correct diagnosis is important because the chemotherapy treatment for GS is different from the commonly incorrect diagnosis of NHL.

Introduction

* Granulocytic Sarcoma (GS) (chloroma) is a rare extramedullary manifestation of acute myeloid leukemia (AML).
* Typically GS manifests in the skin, soft tissues, lymph nodes and bones.1
* In less than 7% of all cases the tumor is found in the small intestines.1
* GS is a more difficult version of AML to manage as there is no way to assess response to chemotherapy.
* If GS goes undiagnosed or untreated, 90% of GS’s will progress to AML.2
* Our patient had no hematologic or bone marrow involvement, his only symptom was a recurrent small bowel obstruction.
* GS is usually misdiagnosed as a malignant lymphoma until appropriate staining is performed due to the similarities under microscopy.2
* Important tumor markers to stain tissue for to make a diagnosis of GS are: myeloperoxidase, lysozyme, c-kit (CD 117), CD43, CD34, CD68, vimentin being positive and cytokeratin, CD3, CD20, and S100 being negative.
* GS is treated as if the patient has AML to achieve best survival rates.3

Discussion

* Typically patient’s in primary GS’s are asymptomatic and go undiagnosed in up to 50%.7
* Tumors respond well to typical AML treatments which include: Daunorubicin + High Dose Cytarabine (3days + 7days).
* But due to the difficulty of monitoring, due to the lack of bone marrow involvement. Tumors usually recur in about 23% of cases and often spread to the bone marrow in <1 year.3,7
* Currently our patient is alive and well greater that 2.5 years out from resection and chemotherapy.

Conclusion

* Granulocytic Sarcomas are rare tumors and even more rare to present as a primary small intestine tumor.7
* Surgical resection is not enough when treating GS and adjuvant chemotherapy aimed at treating AML is required to increase chances of remission.
* Appropriate staining of tumors is needed to make appropriate diagnosis to avoid incorrect treatment.

References:

Figure 1: CT scan of mass seen in small bowel

Figure 2: Microscopic examination of tumor


Designed by Michelle Snavely.