Case 9

Myeloid Sarcoma of small intestine

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Background

Myeloid sarcoma is a extramedullary tumour comprised of myeloblasts and immature myeloid cells. Myeloid sarcoma was previously known as Chloroma due to gross greenish appearance identified in some lesions. Other synonyms for this neoplasm are extramedullary myeloid tumor and granulocytic sarcoma (GS). According to World Health Organization classification of haematopoietic tumours myeloid sarcoma divided into two major categories. The more common form is GS, composed mainly of myeloblasts, neutrophils, and myeloid precursors. The less common form is known as monoblastic sarcoma. It can involve any organ of the body however skin, bone, spine, lymph nodes, soft tissue and genitourinary tract involved more commonly. Gastrointestinal tract is uncommon site of involvement (7%) and small bowel (10%) is the most common region affected in this system. In less than 10% cases multiple anatomical sites can be involved. The male to female ratio is1.2:1 and median age of involvement is 56 years. Clinically it may be initial presentation or a relapse of acute myeloid leukemia (AML). It may also represent blastic transformation of myelodysplastic syndromes or myeloproliferative neoplasms as well as in patients with no known haematological disorders. Clinical symptoms are epigastric pain, chronic anemia and acute upper gastrointestinal bleeding, perforation and small bowel obstruction. AML most frequently associated with granulocytic sarcoma is acute myeloblastic leukemia with maturation (French-American-British [FAB] M2). Most common cytogenetic abnormalities associated with GS is t(8;21) translocation and less frequently the inv(16) type.

Case report

60 year old male presented with abdominal distension and pain of 2 days duration. He also had fever with chills and breathlessness since 1 month. USG Features suggestive of small intestinal obstruction with moderate ascitis. CECT shows Diffuse mass (26mm thick) encasing aorta from annulus upward, extending into right atrium; suggestive of malignant tumor. Multifocal eccentric bowel wall thickening with one of the loops in the pelvis causing complete and another one partial obstruction. Jejuno-jejunal intussception is also seen. Diffuse infiltrative mass lesion infiltrating the visualized heart walls described above. Small nodule in superior segment of right lower lobe of lung. Findings were suggestive of disseminated lymphoma. Exploratory laprotomy and resection was performed.

We received segment of jejunum measuring 15x5 cms. External surface was greyish white with few congested areas. On cutting open a ulceroinfiltrative growth was present. The growth measured 5x3 cm. The cut section of tumor was greyish white and firm and tumor was seen reaching upto serosa. A small nodule measuring 1.5x0.7 cm was seen situated 2 cm away from growth. The remaining mucosa appears unremarkable. Microscopy revealed small intestine showing ulceration of mucosa with areas of necrosis and a tumor which extending up to serosa. Tumor was comprised of diffusely arranged discohesive cells without any pattern. The cells were medium to large with scant to moderate amount of eosinophilic cytoplasm. The medium cells have round to oval nuclei with inconspicuous nucleoli. The large cells have irregular and embryo like vesicular nuclei with prominent nucleoli. Also seen are binucleate and multinucleate tumor giant cells. Mitotic activity was brisk. Sections from the adjacent intestine revealed tumor deposits but cut margins were free of tumor. From above features we come to the diagnosis of High grade non Hodgkin lymphoma of small intestine with two differentials one was pleomorphic variant of plasmablastic lymphoma and second was anaplastic large cell lymphoma.

Conclusion

The wide variety of manifestations of myeloid sarcoma makes the diagnosis very difficult, especially in aleukemic cases. It may be mistaken for diffuse large cell lymphoma of small bowel. Gastrointestinal involvement is very unusual and we must maintain a high index of suspicion. Immunohistochemical techniques are needed for accurate diagnosis. The prognosis of MS is variable but seems to be somewhat similar to that of AML. Prompt recognition and early institution of therapy is crucial, and may delay its progression to a systemic disease. Treatment option for this condition includes surgical resection, systemic chemotherapy and allogeneic bone marrow transplantation.