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Myeloid Sarcoma Presenting as Rectosigmoid Tumour: A Case Report

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ABSTRACT

Myeloid Sarcoma (MS) or extramedullary myeloid tumor is a rare tumor affecting the skin, lymph node, and any part of the body except the bone marrow. MS may present differently depending on the site of involvement. A 61-year-old male with a known case of chronic myeloid leukemia presented to the emergency department of a district hospital with per rectal bleeding for two days. Examination revealed haematochezia with no evidence of rectal tumor. Hyperleukocytosis was noted from the blood investigation. Colonoscopy was performed, which revealed the rectosigmoid tumor. Histopathological examination confirmed the diagnosis of myeloid sarcoma. The patient was subsequently referred to a hematologist in a tertiary hospital for further care. MS may present a diagnostic challenge as it may present nonspecific symptoms depending on the site of involvement. Awareness of this condition is essential; thus, diagnosis and treatment can be initiated earlier.

Introduction

Myeloid Sarcoma (MS) is a tumor of immature myeloid cells or myeloblast that occur in the area other than the bone marrow. It was first described by Burns in 1811. In 1853, King reported a "green-colored tumor", naming the tumor as chloroma owing to the production of myeloperoxidase by the tumor [1]. In 1967, Rappaport proposed the term "granulocyte sarcoma" to describe any extramedullary manifestation of Acute Myeloid Leukemia (AML) [1-3]. In 2008, the World Health Organization (WHO) defined MS as a tumor mass with myeloid blasts that can be mature or immature at any anatomical site other than bone marrow [1]. It is also known as an extramedullary myeloid tumor. MS is most commonly present over the skin, lymph node, bone, and soft tissue. Pileri et al. explored

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92 patients who reported that skin was the most typical site of MS manifestation [4]. Rarely, it may present as a tumor over the gastrointestinal tract. We present a case of MS presented as the rectosigmoid tumor.

Case Presentation

A 61-year-old male with underlying Chronic Myeloid Leukemia (CML) presented to the emergency department with a two-day history of generalized abdominal pain associated with haematochezia, weight loss, and appetite loss. According to his family members, he also could not communicate properly and appear confused for the past two days. The patient was previously diagnosed with chronic myeloid leukemia and was on regular tablet Imitinab 400mg twice daily; however, he defaulted his follow-up and medication for the past two years.

Upon assessment, his Glasgow Coma Scale (GCS) was E4V1M5, with his pupil bilaterally reactive to light. His blood pressure equaled 160/85 mmHg, his heart rate was 82 beats per minute, and his oxygen saturation was 98% under nasal prong oxygen. His abdomen was soft without guarding or tenderness. The spleen was enlarged. Hematochezia was noted during per rectal examination but no rectal mass. No hemorrhoid or rectal ulcer was visualized during proctoscopy. Neurological examination was normal other than the power score of 4 in both bilateral upper and lower limbs. Blood tests showed hyperleukocytosis. The white blood cell count was measured to be 382.5 x 10^9/L. Other laboratory test results presented the following data: haemoglobin: 8.6 g/dL, platelet: 402 x 10^9/L, urea: 6.9 mmol/L, and creatinine: 180 umol/L.

Urgent Peripheral Blood Film (PBF) revealed hyperleukocytosis with neutrophils predominant (Figure 1). Left shift maturation was noted with 5% blast. No Auer rod was noted, and no significant dysplastic changes were detected. Features are suggestive of chronic myeloid leukemia in the chronic phase. However, confirmation can only be obtained with bone marrow aspirate and trephine biopsy.

Plain Computer Tomography (CT) was performed (Figure 2). Acute intraparenchymal hemorrhage at the left frontal lobe with intraventricular extension, mass effect, midline shift, and early hydrocephalus was noted. Neurosurgical team input was sought and subsequently treated as spontaneous intracerebral haemorrhage without active surgical intervention.

He was admitted to the ward and started on tablet hydroxyurea 500 mg daily for hyperleukocytosis. We proceeded with colonoscopy given persistent per rectal bleeding. Colonoscopy revealed a non-obstructing rectosigmoid tumor 5 cm to 20 cm from the anal verge (Figure 3). Biopsy was taken and provided for Histopathology Examination (HPE). Carcinoembryonic Antigen (CEA) tumor marker was taken, i.e., normal (1.53 ng/mL). Contrast-Enhanced Computer Tomographic (CECT) of thorax, abdomen, and pelvic was arranged. The patient was subsequently referred to Haematologist, and the Haematologist took over care before further imaging could be undertaken. The histopathology examination result revealed myeloid sarcoma (metastatic myeloid sarcoma) two weeks later.

Discussion

MS may manifest itself differently depending on the site of involvement. Depending on the size of the tumor and location of involvement, the patient may present with compressive or obstructive symptoms [3]. However, up to 50% of patients with MS remain asymptomatic [5]. Our patient presented with per rectal bleeding, which prompted us to proceed with colonoscopy. However, if the patient did not present with haematochezia, he will most likely be treated as CML in blast crisis. No urgent colonoscopy or imaging will be conducted, and the diagnosis of MS may be missed or delayed.

MS may be presented in 4 clinical settings; as de novo preceding the onset of myeloid leukemia by months or years; as the earliest manifestation of blastic transformation in the patient with the myelodysplastic or myeloproliferative disorder; as relapse in a patient previously treated for leukemia; and as a clinical manifestation in patients with known AML [1, 3]. Approximately 2.5% and 9.1% of the patients had MS concurrently with AML [1, 6].

MS may be diagnosed by histopathology, immunohistochemistry, and immunophenotyping. Morphologically, myeloid cell infiltration appears as cancer cell proliferation can be detected. These cancer cells may include myeloblast, monoblasts, or promyelocytes. Ki67 (proliferation index) is usually high and between 50%-95% [1-3]. The WHO 2008 classification stated that myeloperoxidase, chloroacetate, and nonspecific esterase should be included in cytochemical stains [1]. Despite being the most sensitive markers for MS, CD43 and lysozyme are not specific markers. Myeloperoxidase may be used to differentiate MS from lymphoma; it is expressed in 66%-96% of all cases [1].

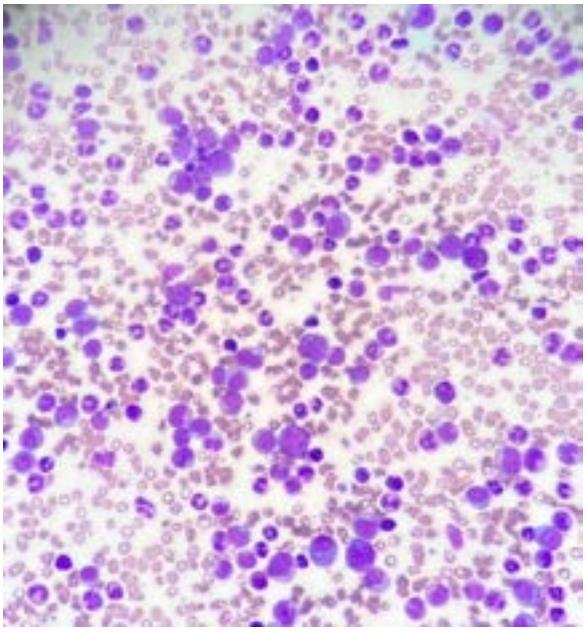

Figure 1. Peripheral blood film viewed under a microscope (40x magnification) indicated hyperleukocytosis

The treatment options for MS include systemic treatment (chemotherapy), local treatment (radiotherapy & surgery), bone marrow transplantation, and targeted therapy. In general, surgery is unnecessary, except to relieve the obstruction or compression caused by the tumor and for diagnostic purposes. Prognosis of the patient with MS varied. Lan et al. reported that patients with MS with CML or myelodysplastic syndrome had a poorer outcome than MS with AML [7]. Systemic chemotherapy treatment was associated with better results.

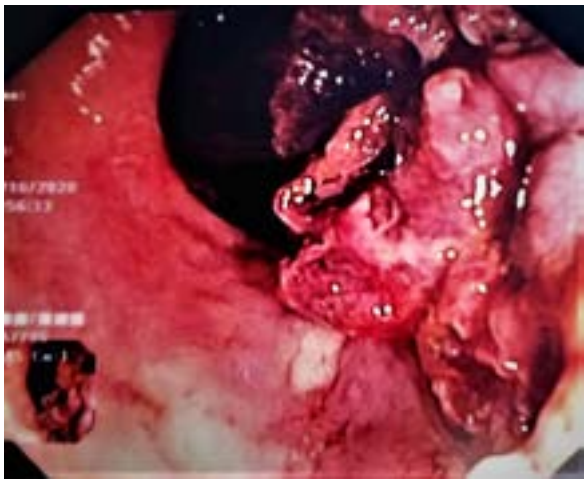

Figure 3. Non-obstructing tumor at rectosigmoid with no evidence of active bleed

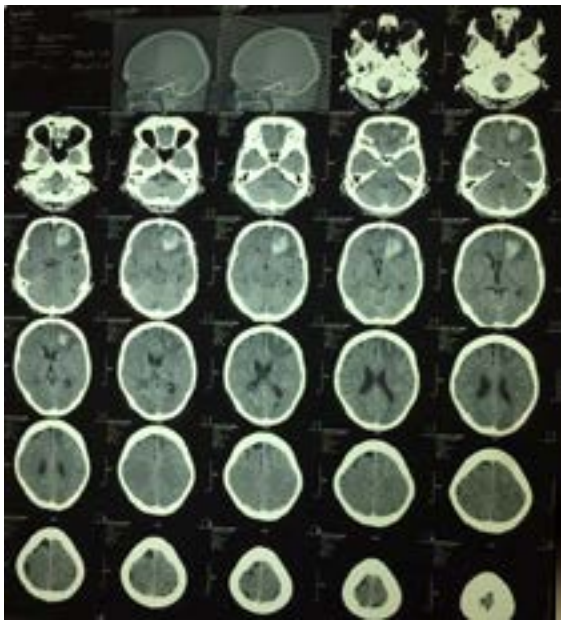

Figure 2. CT brain showed acute intraparencyhmal bleed at the left frontal lobe

Two critical issues are raised in our case report. First and foremost, should all patients diagnosed with AML and CML undergo screening for MS by doing regular endoscopy procedures and imaging? As mentioned before, up to 50% of patients with MS remain asymptomatic. Unless the patient presented with symptoms specific to anatomical location, such as obstruction, compression, or per rectal bleeding, no further imaging or endoscopic procedure will be arranged to screen for MS. MS might be under-diagnosed or under-reported. Universal recommendation for at least yearly imaging and endoscopy will reduce the odds of missing this condition. However, it brings us to the second issue; the cost. Due to its rarity and variable prognosis, the yearly recommendation for endoscopy and imaging might not be cost-effective. Further study may be needed before we can make any sound recommendation.

Conclusion

Due to the nonspecific symptom, MS affecting the gastrointestinal tract may present a diagnostic challenge to clinicians. It remains debatable whether routine endoscopy and imaging should be performed in all patients with the myeloproliferative disorder and myeloid leukemia to screen for MS. Further study may be required before any conclusions could be made.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this article. Informed consent has been obtained from the patient for the publication.

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Conflict of interest

The authors declared no conflict of interest.

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