

Case report**INTESTINAL MYELOID SARCOMA**

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Abstract

This study presents a rare presentation of extramedullary myeloid tumor , affecting terminal ileum in a 25 year old married female patient , managed acutely as acute appendicitis , which was proved histologically to be due to myeloid sarcoma in form of a tumour mass of myeloblasts or immature myeloid cells infiltrating terminal ileum .

Key words: Acute myeloid leukemia , Extramedullary myeloid tumor , myeloid sarcoma , granulocytic sarcoma, chemotherapy .

1. Case presentation:

A 25 years old female , married , Who was before medically free , admitted through emergency with progressive right iliac fossa pain , decrease oral intake , on-off fever , and constipation lasted about 20 hours with no urinary symptoms . Physically was looking ill ,in pain , low grade fever , local right iliac fossa tenderness and positive rebound . Chest was clear .CBC, KFT , LFT , Urine analysis , Chest XRAY were Normal .The patient prepared and shifted to theatre as clinical suspicion of acute appendicitis . on opening firm mass involving appendix ,mesocolon , and terminal ileum. Appendectomy , 34 cm of terminal ileum segment , fatty tissue of 10 x 14 cm were resected .The histology report was involvement of appendix ,mesocolon , and terminal ileum by myeloid sarcoma , transmural infiltration with four matted lymph nodes ,with CD68-KP1 , myeloperoxidase (MPO), CD117, CD99 were positive confirmative markers . After full recovery the patient referred to our oncology department for further management. Concurrent acute myeloid leukemia ruled out, Bone marrow aspirate and biopsy were normal with no blasts, Abdominal Ultra sound revealed minimal free fluid in abdomen and pelvis, CT chest , abdomen and pelvis revealed no masses .

2. Result:

this patient was ideally managed with AML conventional chemotherapy , went through induction 3+7 protocol (Daunorubicin of 3days + cytarabine of 7 days) and 3 consolidation (high dose cytarabine) , with regular follow up during and after chemotherapy where clinical assessment and investigational evaluation of peripheral morphology , bone marrow study , and CT scan ruled out medullary and extra medullary AML relapse , The patient doing well since surgery about 18 months ago , and kept under regular follow up .

3. Discussion:

myeloid sarcoma (chloroma, granulocytic sarcoma or extramedullary myeloid tumor) is an extramedullary manifestation of acute myeloid leukemia . Myeloid sarcoma is a rare disease and is most often found either concurrently or following a previously recognized AML, It may also occur as an isolated leukemic tumor or precede the appearance of blood or bone marrow disease⁽¹⁻³⁾. MS is reported in 2-8% of patients with AML either as a single or as a multifocal tumor. It can predate AML by months or years in approximately a quarter of cases, appear concomitantly with AML in 15-35% of cases, or occur after the diagnosis of AML in up to 50% of cases⁽⁴⁾. The age of patients at MS presentation is highly variable, Biopsy proven MS is most

commonly reported in the skin, bone and lymph nodes. It can however involve many other body sites with reported cases in the central nervous system, oral and nasal mucosa, breast, genitourinary tract, chest wall, pleura, retro peritoneum, gastrointestinal tract and testis⁽⁵⁾, the most common signs and symptoms associated with the myeloid tumor are compression signs accompanied by severe pain and abnormal bleeding, Computerized tomography (CT) and magnetic resonance imaging (MRI) are often used for tumor localization, The most common positive markers in paraffin CD68/KP1, MPO, CD117, CD99, CD68/PG-M1, lysozyme, CD34, TdT, CD56, CD61, CD30, glycophorin and CD4^(6,7). Myeloid sarcoma is associated with superior event-free survival and overall survival compared with acute myeloid leukemia⁽⁸⁾, The current recommended treatment regimen in patients presenting with isolated MS or MS presenting concomitantly with AML is conventional AML-type chemotherapeutic protocols⁽⁹⁾

References

1. Kasahara S, Tsurumi H, Hara T, et al. *Idiopathic myelofibrosis developing isolated granulocytic sarcoma with der(1;7)(q10;p10) after splenectomy and finally transforming to acute myelogenous leukemia. Leuk Lymphoma. 2000;39:427–433.*
2. Cankaya H, Ugras S, Dilek I. *Head and neck granulocytic sarcoma with acute myeloid leukemia: three rare cases. Ear Nose Throat J. 2001;80:224–226.*
3. Suzer T, Colakoglu N, Cirak B, et al. *Intracerebellar granulocytic sarcoma complicating acute myelogenous leukemia: a case report and review of the literature. J Clin Neurosci. 2004;11:914–917.*
4. Maeng H, Cheong JW, Lee ST, et al. *Isolated extramedullary relapse of acute myelogenous leukemia as a uterine granulocytic sarcoma in an allogeneic hematopoietic stem cell transplantation recipient. Yonsei Med J. 2004;45:330–333.*
5. Pileri SA, Ascani S, Cox MC, et al. *Myeloid sarcoma: clinico-pathologic, phenotypic and cytogenetic analysis of 92 adult patients. Leukemia. 2007;21:340–350.*
6. Hani Al-Khateeb, Ahmed Badheeb, Husam Haddad, et al. *Myeloid Sarcoma: Clinicopathologic, Cytogenetic, and Outcome Analysis of 21 Adult Patients. Volume 2011 (2011), Article ID 523168, 4 pages*
7. Markoc F, Bozdogan N, Yükrük FA, et al. (2010) *Granulocytic sarcomas: Difficulties in diagnosis. Tumori 96:149–153.*
8. A. Tsimberidou, H. M. Kantarjian, S. Wen et al., “Myeloid sarcoma is associated with superior event-free survival and overall survival compared with acute myeloid leukemia,” *Cancer*. 2008 Sep 15;113(6):1370-8. doi: 10.1002/cncr.23691. *Cancer*, vol. 113, no. 6, pp. 1370–1378, 2008
9. Batia Avni, MD and Maya Koren-Michowitz, MD *Myeloid Sarcoma: Current Approach and Therapeutic Options Ther Adv Hematol. Oct 2011; 2(5): 309–316.*