World Cancer Research Journal WCRJ 2019; 6: e1429

MYELOID SARCOMA OF THE SPLEEN: REPORT OF A RARE CASE WITH REVIEW OF LITERATURE

M. RAMEZANI[1], K. SEIDI[2], F. AMIRIAN[1], M. SADEGHI[3]

[1]Molecular Pathology Research Center, Imam Reza Hospital, Kermanshah University of Medical Sciences, Kermanshah, Iran
[2]Students Research Committee, Kermanshah University of Medical Sciences, Kermanshah, Iran
[3]Medical Biology Research Center, Kermanshah University of Medical Sciences, Kermanshah, Iran

Abstract – *Objective:* *Myeloid sarcomas (MSs) are rare tumors of myeloid blasts and immature cells. Rare cases of splenic involvement are on record. We report a case of MS in a young female which is confirmed on pathologic examination of the spleen.*

Patients and Methods: *A 20-year-old female was admitted with the chief complaints of abdominal pain, fever, anorexia, and dyspnea. She was a known case of acute myelogenous leukemia (AML). The spiral computed tomography scan without contrast showed a massive splenic enlargement. The ultrasound examination showed liver and spleen were larger than normally measured 190 and 210 mm, respectively, with normal parenchymal echo.*

Results: *The patient underwent surgery with clinical suspicion of MS. The specimen consisted of the red spleen with several infarcted areas totally measuring 22x14x10 cm and weighing 1885 g. The pathology report was splenic infarction with myeloid sarcoma.*

Conclusions: *The risk of misdiagnosis is high in the pathologic exam, especially in non-leukemic forms and low clinical suspicion. It is important to consider this entity in the differential diagnosis of splenomegaly or splenic mass in any age group.*

KEYWORDS: *Myeloid sarcoma, Chloroma, Granulocyic sarcoma, Spleen, Case report.*

INTRODUCTION

Granulocytic sarcomas, chloromas or myeloid sarcomas (MS) are rare tumors originating from myeloid blasts and immature cells and most data in the literature are from case reports and case series[1-3]. It is estimated with an incidence of up to 9% of acute myelogenous leukemias (AMLs)[4]. Some researchers stated an incidence of as low as 0.8% of AMLs[5]. This tumor is reported in bone, periosteum, soft tissue, lymph node, skin, and other organs[6]. The most common site of presentation is demonstrated in connective/soft tissue followed by skin/breast[5]. The involvement of the liver, spleen, brain, and other organs is also on the report[7]. Herein, we report a case of MS in a young female which is confirmed on splenectomy pathologic diagnosis according to morphology and clinical data.

CASE REPORT

A 20-year-old female was admitted on 19th January 2017 at the Oncology Department with the chief complaints of abdominal pain, fever, anorexia, and dyspnea. She was a known case of AML since 2 months before. On 6th November 2016, bone marrow aspiration and trephine biopsy were carried out and the pathologist reported: Hyper cellular marrow (cellularity 100%) containing 50% atypical monocytoid cells suggestive for acute myelogenous leukemia (FAB-M5). At the same time, a computed tomography (CT) scan of the lung and mediastinum demonstrated massive effusion with collapse and consolidation in the left hemithorax and shift of the heart and mediastinum to the right. There was segmental atelectasis in the left para cardiac region. The spiral CT scan without

Corresponding Author: Masoud Sadeghi, MSc; e-mail: sadeghi_mbrc@yahoo.com

contrast showed a moderate amount of fluid in the left pleural space and parenchymal opacity in the basal segment of the left lower lobe and lower segment of lingual suggestive for the infectious process. The massive splenic enlargement was noted. The patient was diagnosed with left pulmonary emboli and chest tube indwelled. The ultrasound examination at the same time reported: gall bladder, kidneys, urinary bladder, endometrium, and ovaries were within normal limits. Few lymph nodes in porta hepatis measuring 20 x 9 mm were noted. The liver and spleen were larger than normally, measuring 190 and 210 mm, respectively, with normal parenchymal echo. The patient received chemotherapy. Bone marrow aspiration and trephine biopsy on 19th December 2016 were normocellular marrow with 5-10% monocytoid cells and less than 5% of blasts. Finally, on 19th January 2017, the patient underwent splenectomy with clinical suspicion of MS. The specimen consisted of the red spleen with several infarcted areas totally measuring 22x14x10 cm and weighing 1885 g. The pathologist reported: compatible with splenic infarction with leukemic involvement (granulocytic sarcoma; Figures 1, 2, and 3). Isolated lymph node showed lymphoma–leukemia involvement. The patient achieved relief of symptoms after splenectomy and discharged with a prescription. The patient was in good condition in the last admission for the removal of the right porto-jugular catheter on 24th April 2018.

DISCUSSION

MS can occur in a wide range of ages from 1 to 81 years[3]. The most common presentation is compression effect, pain, and abnormal bleeding[3]. Wang and Li[8] reported 39 cases of myeloid sarcoma with a mean age of 33.4 years; one of them was originating from the spleen with clinical expression of massive splenomegaly and traumatic rupture. Most of the cases showed poor prognosis. Goyal et al[5] reported 746 cases of myeloid sarcoma with a median age of 59 years. They considered cases of lymph node and splenic involvement in one prognostic group and considered them as poor prognosis along with cases of the nervous system, soft tissue, mediastinum, and bone. Patients with less than 70 years had a better prognosis than the older ones. Median overall survival was worse in men, blacks, and poor prognostic sites[5]. Reports of cases with unusual age and presentation are on record. Kim and Kim[9] reported myeloid sarcoma of the breast with hamartoma-like features in ultrasound examination; finally biopsy confirmed the diagnosis. Marwah et al [10] reported myeloid sarcoma as a large temporal bone mass in a child with Down syndrome as the initial manifestation of the disease. They emphasized on diverse sites of presentation and the need for rapid diagnosis to achieve better survival. Others[11] confirmed this idea with a report of the mass in paranasal sinus with presentation as epistaxis. Moreover, Mitkowski and Gil[12] reported the eyelid of a 36 years old woman. Tokunaga et al[13] reported a man with pancreatic mass as the manifestation of Granulocytic sarcoma that showed complete remission after hematopoietic stem cell transplantation. Rao *et al* [14] reported a case of non-leukemic granulocytic sarcoma of the spleen in an infant and emphasized early diagnosis. One study[15] reported a 58 years old woman with acute onset of fever and abdominal pain, which was diagnosed as acute myelogenous leukemia, FAB M2 (French-American-British classification). Core needle biopsy of the splenic mass confirmed the diagnosis of granulocytic sarcoma; however, de-

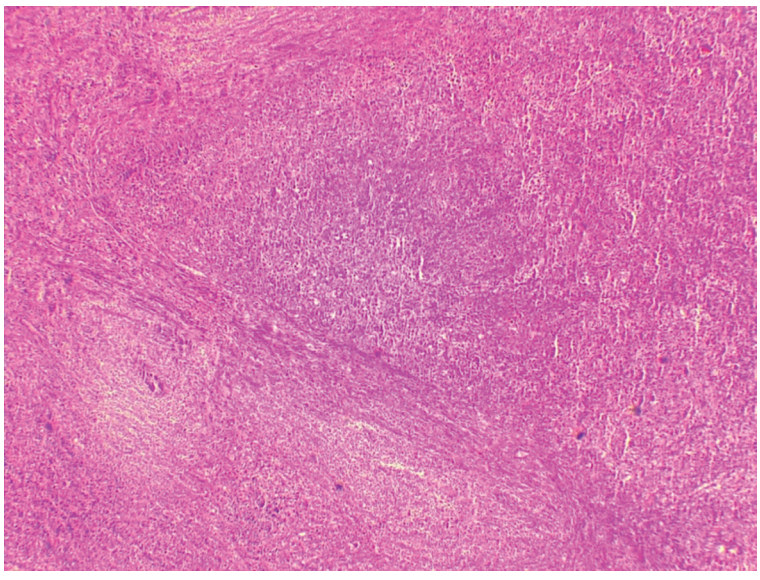

Fig. 1. Diffuse infiltration of tumor cells with high nucleo-cytoplasmic ratio in the spleen, myeloid sarcoma of the spleen. Hematoxylin-Eosin (HE) staining (X40 magnification).

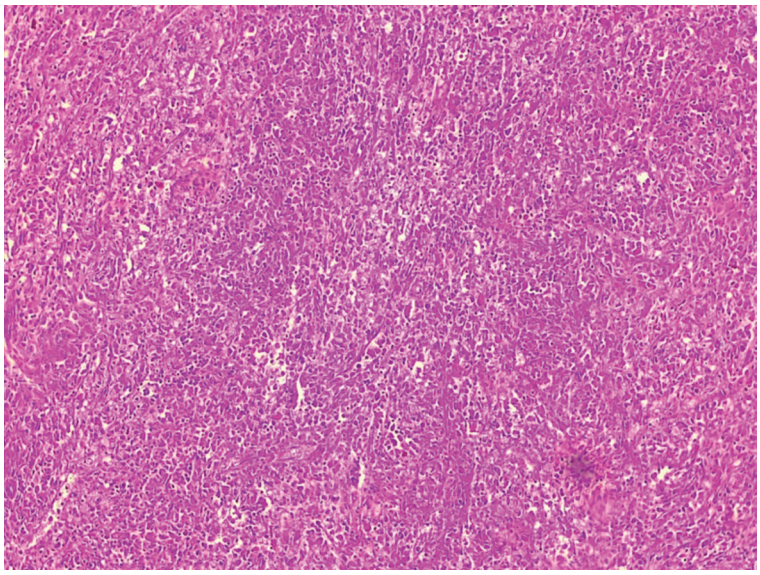

Fig. 2. Diffuse infiltration of tumor cells with high nucleo-cytoplasmic ratio in the spleen, myeloid sarcoma of the spleen. Hematoxylin-Eosin (HE) staining (X100 magnification).

layed targeted therapy culminated in mortality. The authors emphasized on early diagnosis and treatment of solitary splenic mass in a patient with acute leukemia.

The risk of misdiagnosis is high in the pathologic examination, especially in nonleukemic forms and low clinical suspicion[16]. In the experience of Neiman et al[6] on 61 biopsies, most tumors were originally diagnosed as lymphoma due to poor differentiation. Histopathology and immunohistochemistry studies are helpful for correct diagnosis[17,18]. A research[19] reported 25 cases of myeloid sarcoma. The most common histopathology pattern was the diffuse arrangement of pleomorphic cells with irregular nuclei, prominent nucleoli, and abundant eosinophilic cytoplasms. Systemic chemotherapy, surgery, radiotherapy, hematopoietic stem cell transplantation, and targeted therapy are different treatment modalities that are indicated according to the patient's condition, presentation and stage of the disease[20].

CONCLUSIONS

Myeloid sarcoma is rare in the spleen and the risk of misdiagnosis is high in the pathologic exam, especially in non-leukemic forms and low clinical suspicion but it is important to consider this entity in the differential diagnosis of splenomegaly or splenic mass in any age group. Early diagnosis and appropriate treatment may result in better survival.

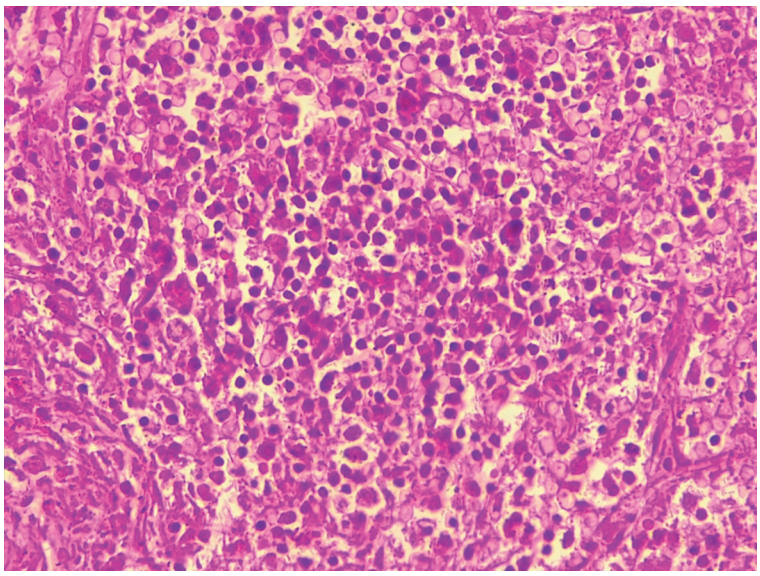

Fig. 3. Diffuse infiltration of tumor cells with high nucleo-cytoplasmic ratio in the spleen, myeloid sarcoma of the spleen. Hematoxylin-Eosin (HE) staining (X400 magnification).

Declaration of patient consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Acknowledgment:

The authors thank Mrs. Sholeh Akradi for her support in providing data. The authors also would like to thank the Clinical Research Development Center of Imam Reza Hospital for Consulting Services.

Conflict of Interest:

There are no conflicts of interest.

REFERENCES

1. Antic D, Perunicic Jovanovic M, Knezevic D, Ostojic S, Elezovic I. Progressive course of isolated splenic myeloid sarcoma. Cent Eur J Med 2013; 8: 499-502.
2. Roby BB, Drehner D, Sidman JD. Granulocytic sarcoma of pediatric head and neck: an institutional experience. Int J Pediatr Otorhinolaryngol 2013; 77: 1364-1366.
3. Avni B, Koren-Michowitz M. Myeloid sarcoma: current approach and therapeutic options. Ther Adv Hematol 2011; 2: 309-316.
4. Ohanian M, Faderl S, Ravandi F, Pemmaraju N, Garcia-Manero G, Cortes J, Estrov Z. Is acute myeloid leukemia a liquid tumor? Int J Cancer 2013; 133: 534-543.
5. Goyal G, Bartley AC, Patnaik MM, Litzow MR, Al-Kali A, Go RS. Clinical features and outcomes of extramedullary myeloid sarcoma in the United States: analysis using a national data set. Blood Cancer J 2017; 7: e592.
6. Neiman RS, Barcos M, Berard C, Bonner H, Mann R, Rydell RE, Bennett JM. Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases. Cancer 1981; 48: 1426-1437.
7. Mei KD, Lin YS, Chang SL. Myeloid sarcoma of the cheek and the maxillary sinus regions. J Chin Med Assoc 2013; 76: 235-238.
8. Wang HQ, Li J. Clinicopathological features of myeloid sarcoma: Report of 39 cases and literature review. Pathol Res Pract 2016; 212: 817-824.
9. Kim SJ, Kim WG. Sonographic features of a myeloid sarcoma of the breast as a relapse of acute myeloid leukemia after stem-cell transplantation: a case report. Am J Case Rep 2019; 20: 612-619.
10. Marwah N, Bhutani N, Budhwar A, Sen R. Isolated myeloid sarcoma of the temporal bone: As the first clinical manifestation of acute myeloid leukemia in a patient of down's syndrome. Int J Surg Case Rep 2019; 58: 77-80.
11. Dewan K, Baird JH, Shires CB. An unusual cause of epistaxis: paranasal sinus myeloid sarcoma. Case Rep Hematol 2019; 2019: 1312630.
12. Mitkowski D, Gil L. Isolated myeloid sarcoma as the first manifestation of acute myeloid leukemia: a case study. Clin Case Rep 2017; 5: 1802-1806.
13. Tokunaga K, Yamamura A, Ueno S, Kikukawa Y, Yamaguchi S, Hidaka M, Matsuno N, Kawaguchi T, Matsuoka M, Okuno Y. Isolated pancreatic myeloid sarcoma associated with t(8;21)/RUNX1-RUNX1T1 rearrangement. Intern Med 2017; 57: 563–568.
14. Rao Y, Wu Y, Dong A, Zhu K, Li W, Cai S, Yang M, Yan J. Disseminated nonleukemic myeloid sarcoma of the spleen with involvement of the liver in an infant. J Pediatr Hematol Oncol 2017; 39: e233-e235.
15. Chen WL, Hsu YJ, Tsai WC, Tsao YT. An unusual case of febrile neutropenia: acute myeloid leukemia presenting as myeloid sarcoma of the spleen. J Natl Med Assoc 2008; 100: 957-959.
16. Singh A, Kumar P, Chandrashekhara SH, Kumar A. Unravelling chloroma: review of imaging findings. Br J Radiol 2017; 90: 20160710.
17. Alexiev BA, Wang W, Ning Y, Chumsri S, Gojo I, Rodgers WH, Stass SA, Zhao XF. Myeloid sarcomas: a histologic, immunohistochemical, and cytogenetic study. Diagn Pathol 2007; 2: 42.
18. Valbuena JR, Admirand JH, Gualco G, Medeiros LJ. Myeloid sarcoma involving the breast. Arch Pathol Lab Med 2005; 129: 32-38.
19. Campidelli C, Agostinelli C, Stitson R, Pileri SA. Myeloid sarcoma: extramedullary manifestation of myeloid disorders. Am J Clin Pathol 2009; 132: 426-437.
20. Almond LM, Charalampakis M, Ford SJ, Gourevitch D, Desai A. Myeloid sarcoma: presentation, diagnosis, and treatment. Clin Lymphoma Myeloma Leuk 2017; 17: 263-267.