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Case Report

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ACUTE ERYTHROID LEUKEMIA (M_6 A): A RARE CASE REPORT

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ABSTACT

Acute erythroid leukemia is a rare form of acute myeloid leukemia. It accounts for 3-4% Of all cases of AML. According to the World Health Organization 2008 classification, it falls under the category of acute myeloid leukemia, not otherwise specified and is further divided into two subtypes: erythroid leukemia (erythroid/myeloid) and pure erythroid leukemia. Currently, erythroleukemia (erythroid/myeloid) is defined as 50% or more erythroid precursors and ≥20% blasts of the non-erythroid cells. By definition, pure erythroid leukemia is composed of ≥80% erythroid precursors. However M_6 A is very rare and here we present a case of 55 year old lady presented with weakness and breathlessness was found to be a erythroid leukemia on bone marrow examination. We present this case for its rarity.

KEY WORDS: AML, M6A,e rythroid leukemia.

INTRODUCTION

Acute erythroid leukemia (AEL) is a rare form of acute myeloid leukemia (AML). It accounts for less than 5% of all AML cases.1 In 1912, Coppelli, described condition he named erithromatosis in a patient who presented with anemia, splenomegaly, foci of erythroblast in liver, spleen, lymph nodes and bone marrow but no circulating blasts.[1]. In 1976, French-American-British (FAB) cooperative group designated AEL as AML-M6 defined as erythroid precursors ≥ 30% and dyserythropoiesis ≥10%.[2,3]. In 1992, Kowal-Vern proposed that Di Guglielmo disease defined as erythroleukemia with ≥ 30% proerythroblasts should be a separate category in the FAB classification because of its worse prognosis compared to Di Guglielmo syndrome.[4] Goldberg (1998) confirmed that GD besides the worse prognosis has distinct clinical, laboratory and cytogentics characteristic[5]. Kowal-Vern et al and Mazzela et al proposed three subsets of AML-M6: M6a (myeloblast-rich erythroleukemia) equivalent to erythroid/myeloid leukemia; M6b (proerythroblast-rich erythroleukemia) equivalent to pure erythroid leukemia and M6c (myeloblast and proerythroblast-rich mixed variant[6,7].

CASE REPORT

55 Yr old female presented to our OPD with generalised weakness, shortness of breath and mild sternal tenderness for 15 days. On examination she was having moderate pallor. She had a history of infusion of two units of blood 3 months back. There was no history of malena, vomiting etc. On systemic examination she was having mild hepatosplenomegaly. Routine Blood tests revealed Hb to be 3.8 gm/dl Total leucocyte count to be 11000/dl and N-48%,L-45%,E-02%,B-00%,atypical cells-05% and retic-1.77%. RBC was normocytic, normochromic with mild anisopoikilocytosis with few target cells. WBC-showed atypical cells. Platelets were reduced to 42000/dl. On peripheral smear it was diagnosed to be bicytopenia with presence of atypical cells. Then she was subjected to **Bone marrow Aspiration cytology from the right** post superior iliac spine. The aspirate revealed a hypercelluar marrow with Erythroid hyperplasia and decrease in maturation (megaloblastoid change) (fig-1). Myelopoiesis showed increased blast cell population constituting 22% of marrow cells with strong MPO +ve. Megakaryopoiesis was suppressed. Lymphocytes and Plasma cells were within normal limit. Mitosis was increased in both erythroid and myeloid blast cell population. Parasites and metastatic cells were not seen (fig-2). From the above finding provisional diagnosis of acute myeloid leukemia (aml m6a) was made.

DISCUSSION

Acute Erythroid Leukemia (AML M6) is a rare form of acute leukemia. It is a distinctive bone marrow disorder characterized by the neoplastic proliferation of the dysplastic erythroid elements mixed with blasts of myeloid origin. The malignancy usually presents in the fifth and sixth decades, but a bimodal peak has been described in conjunction with AML M6.[8] The smaller peak has been noted below 20 years, and a broader peak in seventh decade.[9] Few pediatric cases have also been elucidated.[10] The age group ranged from 11 to 72 years with a mean of 43.8 years. The signs and symptoms of AML M6 are nonspecific and are attributed to the

replacement of bone marrow elements by neoplastic cells. Patients rarely present with symptoms lasting longer than six months, and they are usually diagnosed within1-3 months after the onset of symptoms.[11] In many series, approximately half the cases of AML M6 are therapy-related while secondary leukemias are less frequent (10-15%).[12] Erythroblasts can be differentiated from other lineages by being positive for Periodic Acid Schiff (PAS), showing globular positivity and negative for Myeloperoxidase (MPO) & Sudan Black B (SBB). In flow cytometry analysis, the erythroblasts are positive for CD36 and glycophorin A, but not specific.[13]

The differential diagnoses for AML M6 could be varied and pose diagnostic challenges[14] With regard to erythroleukemia (erythroid/myeloid); MDS (refractory anemia with excess blasts): is a possibility but the blasts account for less than 20%. In AML with MDS related changes, >20 % blasts with multi-lineage dysplasia in >50% cases of the cells in more than 2 lineages supports the diagnosis. AML with increased erythroid precursors (lesser proerythroblasts and basophilic erythroblasts) also comes in this realm. In cases of pure erythroid leukemia, megaloblastic anemia, acute lymphoblastic leukemia (especially in pediatric age group) and lymphomas are often considered as close diagnostic parallels.[15]

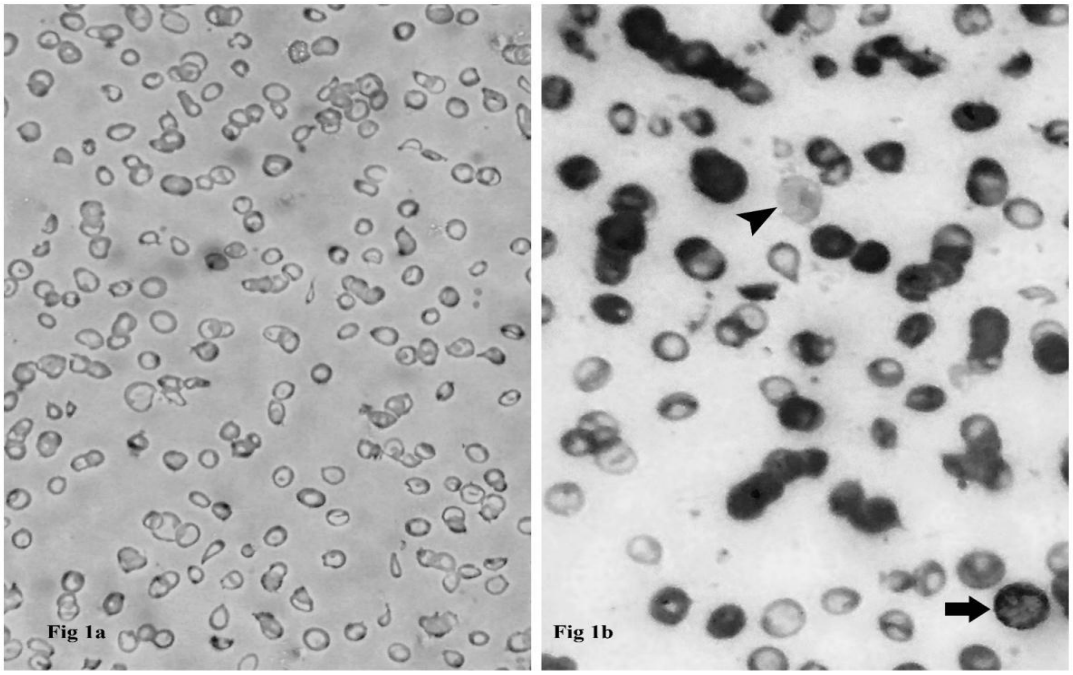

Fig-1: BM HYPERCELLULARITY

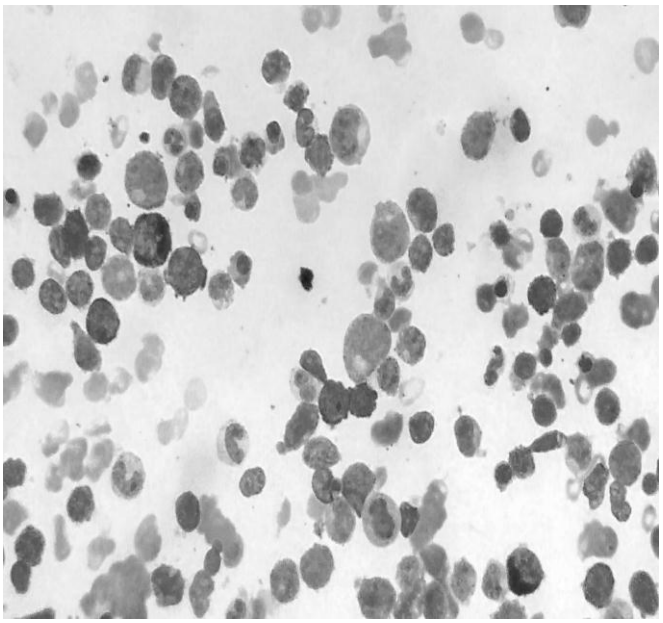

Fig-2: BM MPO POSITIVE MB AND MPO NEGATIVE EB 1

CONCLUSION

Acute erythroid leukemia is a heterogenous disease, which may arise de novo, secondary to MDS or after cytotoxic therapy. Based on the WHO 2008 criteria, AEL becomes diagnosis of exclusion and its frequency is decreased. The differential diagnosis is broad and includes reactive processes or other hematological malignancies. A thorough clinical history, laboratory data, cytochemical and immunophenotypic analysis, genetic and molecular studies are necessary for the correct diagnosis. Additional cytogenetic and molecular studies are required to elaborate our understanding of this disease.

REFERENCES

1. Bain BJ. Di Guglielmo and his syndromes. Br J Haematol. 2003; 120(6): 939-943.
2. Bennett JM, Catovsky D, Daniel MT, et al. Proposals for the classification of the acute leukaemias. French-American-British (FAB) co-operative group. Br J Haematol. 1976; 33(4): 451-458.
3. Kasyan A, Medeiros LJ. Acute erythroid leukemia. Arch Pathol Lab Med. 2010; 134(9): 1261-1270.
4. Kowal-Vern A, Cotelingam J, Schumacher HR. The prognostic significance of proerythroblasts in acute erythroleukemia. Am J Clin Pathol. 1992; 98(1): 34-40.
5. Goldberg SL, Noel P, Klumpp TR, Dewald GW. The erythroid leukemias: a comparative study of erythroleukemia (FAB M6) and Di Guglielmo disease. Am J Clin Oncol. 1998; 21(1): 42-47.
6. Kowal-Vern A, Mazzella FM, Cotelingam JD, et al. Diagnosis and characterization of acute erythroleukemia subsets by determining the percentages of myeloblasts and proerythroblasts in 69 cases. American Journal of Hematology. 2000; 65(1): 5-13.
7. Mazzella FM, Kowal-Vern A, Shrit MA, et al. Acute erythroleukemia: evaluation of 48 cases with reference to classification, cell proliferation, cytogenetics, and prognosis. Am J Clin Pathol. 1998; 110(5): 590-598.
8. Mazzella FM, Alvares C, Kowal-Vern A. The acute erythroleukemias. Clin Lab Med 2000; 20: 119-37.
9. Park S, Picard F, Dreyfus F. Erythroleukemia: a need for a new definition. Leukemia 2002; 16: 1399-401.
10. Batra V V, Karan A S, Saxena R; Erythroleukemia. A clinico-hematological review of 4 cases. In brief reports: Indian Pediatrics 2004; 41: 587-90.
11. Attili SVS, et al. A retrospective study of clinico-hematological and cytogenetic profile of erythroleukemia from South India. Turk J Hematol 2006; 23: 158-63.
12. Domingo-Claros A, Larriba I, Rozman M, et al. Acute erythroid neoplastic proliferations. A biological study based on 62 patients. Hematologica 2002; 87: 148-53.
13. Latif N, Salazan E, Khan R et al. The pure erythroid leukemia: A case report and literature review. Clinical Advances in Hematology and Oncology 2010; 8(4): 283-88.
14. Swerdlow S H, Campo E, Harris N L, Jaffe E S, Pileri S A, et al. Acute myeloid leukemia, not otherwise specified. In: WHO classification of Tumours of Hematopoietic and lymphoid tissues. 4th edition; IARC press, Lyon, France; 2008; 130-9.
15. Khanna R, Nayak DM, Manohar C, Seshadri S, Belurkar SV. Pure erythroleukemia- A case report and literature review. The Internet Journal of Haematology 2011; 7(2): doi: 10.5580/feb.