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A RARE CASE REPORT ON HYPOPLASTIC ACUTE MYELOID LEUKEMIA (H-AML-M5)

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ABSTRACT

Hypocellular acute myeloid leukemia (H-AML) is an infrequent entity. Its frequency ranges between 5-12% of all cases of AML [1,2]. Hypocellular AML is currently defined as AML with a bone marrow cellularity < 20%, although in some earlier reports, cellularity < 40% or 50% was considered to be hypocellular [2,3]. We report a rare case of hypocellular AML-M5 occurring in a 75-year-old man.

KEYWORDS: hypoplastic acute myeloid leukemia, H-AML, AML-M5.

INTRODUCTION

Hypoplastic or Hypocellular Acute Myeloid Leukemia (H-AML) represents a small number of patients diagnosed with myeloid malignancies with a frequency ranging from 5-12% [1,2]. Hypocellular variants of AML almost always have a myeloid phenotype and develop secondary to radiation or chemotherapy [4,5]. Therefore, it mainly affects the elderly. However, H-AML accounts for 5-7% of *de novo* AML[6].

CASE REPORT

A 75-year-old male patient presented with complaints of mild intermittent fever since 20 days along with gradually progressive dyspnea on exertion since last 15 days. There was history of black coloured stools, decreased appetite and weight loss. Patient was diagnosed with pulmonary Koch's 3 years back and had taken anti tubercular treatment for 3 months (Partially treated). There was history of blood transfusion in 2004 (1 unit PCV), in 2014 (2 unit PCV) and in 2016 (2 unit of PCV). On examination, pulse was 94/min, regular, BP 110/70mmHg, Pallor ++, Bony tenderness ++, No lymphadenopathy, clubbing, cyanosis, edema or icterus. On abdominal examination there was no hepatosplenomegaly. Cardiovascular, Respiratory and Central nervous system were within normal limits. No history of chemotherapy or radiotherapy.

Laboratory investigations revealed Hemoglobin–5.5 gm%, total leukocyte count–3050 cells/cumm, differential leukocyte count: 6%Neutrophils, 11% Lymphocytes, 1% Eosinophils and 82% Monocytoid cells, Platelet count- 41,000 cells/cumm, Peripheral blood smear showed a macrocytic picture with moderate anisocytosis and presence of monocytoid cells [Figure 1]. No hemoparasite was seen. The RBC indices were MCV – 112.8fl, MCH – 50.9 pg, MCHC – 45.1g/dl. Blood sugar level, serum protein, blood urea nitrogen, creatinine and serum electrolytes were within the normal range. Urine routine and microscopy was normal.

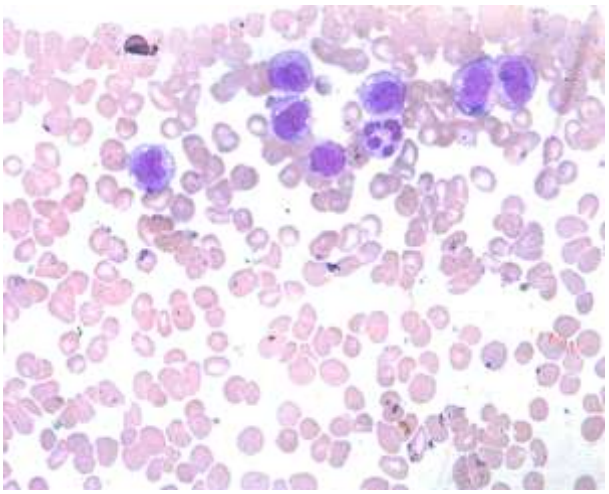

Fig. 1: Peripheral blood smear showing Monocytoid cells and a neutrophil (Leishman stain 40x).

Bone marrow aspiration revealed a dry tap. Imprint and bone marrow trephine biopsy showed features of a hypoplastic marrow with a myeloid to erythroid ratio of 20:1.Erythropiesis was markedly suppressed; Myelopoiesis showed increased blast population of 32% with a monocytoid appearance of folded nuclei,

moderate amount of cytoplasm and inconspicuous nucleoli. Megakaryocytes were markedly reduced. [Fig.2,3]

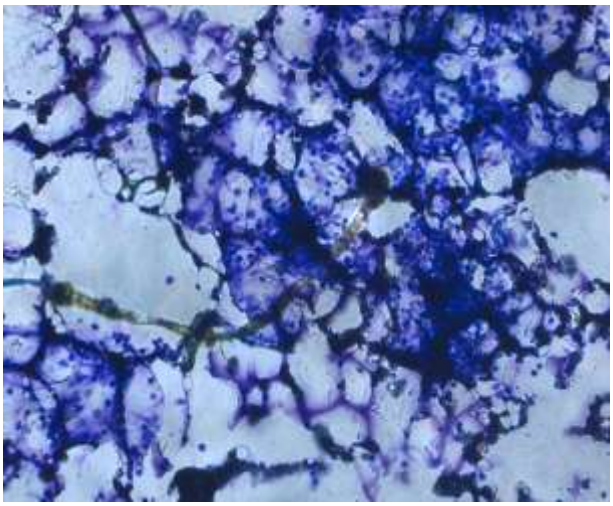

Fig. 2: Bone marrow Aspirate showing features of a hypoplastic marrow [Leishman 10x]

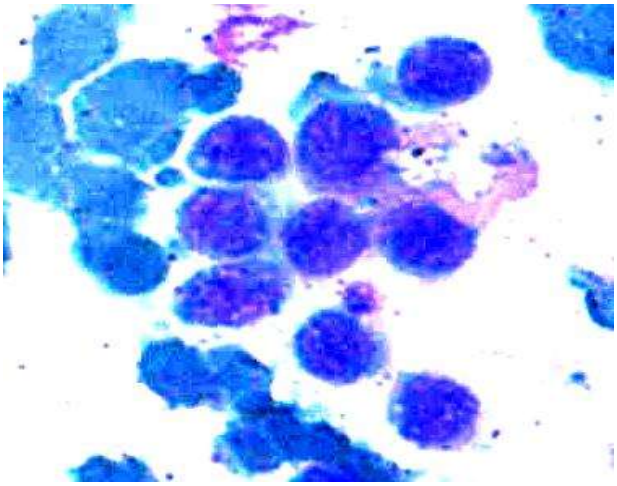

Fig.3: Bone marrow biopsy- showing blasts with moderate amount of cytoplasm, folded nuclei and inconspicuous nucleoli [H&E 40X]

Cytochemically, these blasts were nonspecific esterase positive and myeloperoxidase negative. Flowcytometry on peripheral blood showed CD13, CD33, CD64, HLA-DR and CD117 positivity [Figure 4]

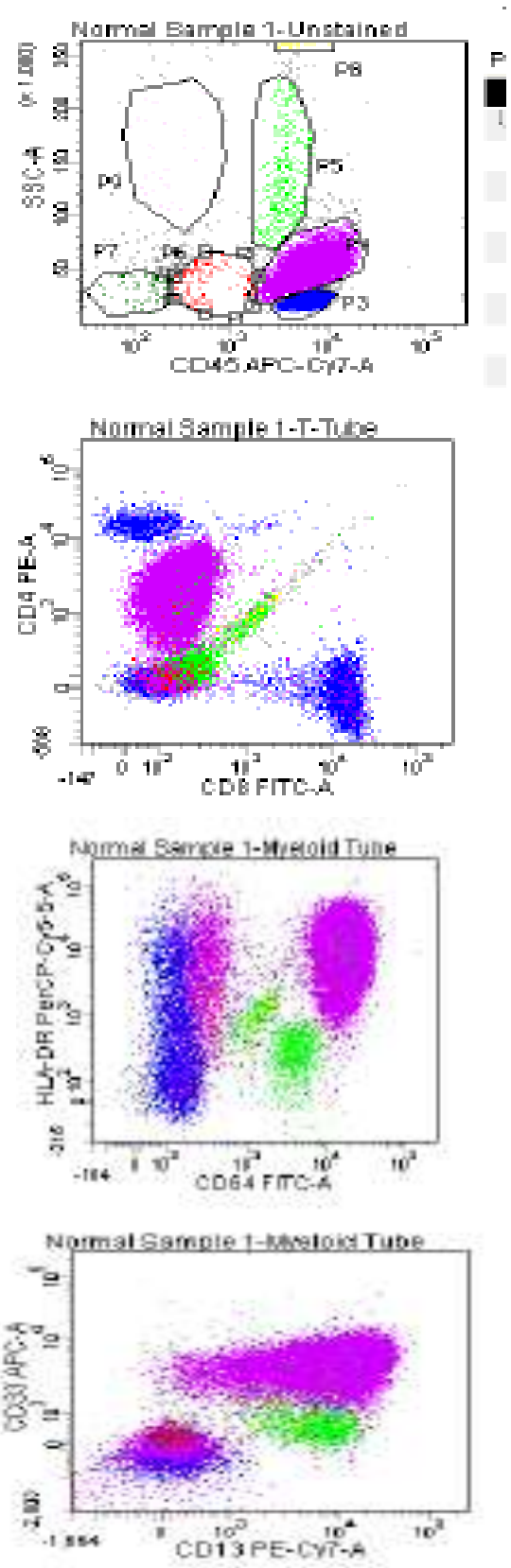

Fig 4: Dot Plot SSC/CD45 gating show cell population in the monocytic windowwith moderate to bright CD45 with positive expression of CD13, CD33, HLA-DR and negative expression of CD8

Based on all these features, a diagnosis of Hypocellular AML-M5 was made.

DISCUSSION

The diagnosis of Hypoplastic AML can be challenging for the hematopathologist since the marrow hypoplasia needs to be differentiated from hypocellular myelodysplastic syndrome and aplastic anemia with marrow dysplasia. Many attempts have been made in the past to establish guidelines for this difficult differential diagnosis. Nagai et al proposed the following diagnostic criteria: (1) pancytopenia with rare appearance of blasts in PB; (2) less than 40% BM hypocellularity; (3) more than 30% blasts in BM and (4) myeloid phenotypes of leukemic blasts by MPO staining and/or immunophenotyping.[4] One such proposal by has been shown in Table1.[4,6,7,8,9,10]

Table 1: Guidelines for diagnosis.

GUIDELINES FOR DIAGNOSIS.

Major recommendations

1. Peripheral blood film: count at least 100 cells wherever possible.
   - Anumerate dysplasia of granulocytes
   - Assess for presence of blasts
2. Bone marrow aspirate
   - Perform a 500 cell differential if possible
   - Examine for dysplasia of erythroid precursors, granulocytes and megakaryocytes
3. Perform an iron stain for ring sideroblast assessment
4. Bone marrow biopsy
   - Assess for cellularity
   - Assess for presence of ALIP (supplement with immunostains for CD34, 117, MPO)
5. Perform a reticulin stain
6. Additional studies
   - Standard cytogenetics/interphase FISH
   - Flow cytometry
   - PNH screening by a sensitive flow or molecular screening technique

Needleman et al.[2] has reported their experience with hypoplastic acute leukemia and suggested that patients with hypocellular bone marrow experience a more indolent course, and can commonly achieve a good response to remission induction therapy. Recently, the beneficial effects of hematopoietic growth factors have been reported in the treatment of hypoplastic AML. It has been observed that chemotherapy may be necessary to maintain remission in hypoplastic AML after hematopoietic reconstitution by granulocyte colony stimulating factor.[11]

CONCLUSION

Hypocellular acute myelomonocytic leukemia is a rare entity, is seen more commonly in older patients as seen in our case. The limited published information about the diagnostic guidelines to be followed in such cases along with the overlap with hypocellular myelodysplastic syndrome and aplastic anemia with marrow dysplasia makes the diagnosis difficult.

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