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RESEARCH ARTICLE

A CASE OF MYELODYSPLASTIC SYNDROME WITH EXCESS BLASTS (MDS-EB-1) PRESENTING WITH BASOPHILIA: A RARE ENTITY AND SHOWING POOR PROGNOSIS

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ABSTRACT

Myelodysplastic syndrome (MDS) with basophilia is a rare condition and has yet to be classified under the 2016 revision to the World Health Organization classification of myeloid neoplasms. However, few reports have described the prognostic significance of basophilia in MDS. Here, we report a case of a 60 year old male who was admitted to the hospital with complains of generalised body weakness, easy fatiguability and breathlessness on mild exertion for one month. Initial investigations showed features of pancytopenia with basophil 12% on complete blood count (CBC). Reticulocyte count was less than 0.5%.Bone marrow aspiration (BMA) showed myeloid and erythroid series cells with features of dysmyelopoiesis and severe degree of dyserythropoiesis. Quadrinucleate, pentanucleate and gigantoblasts forms of erythroblasts were also noted. These findings led to the diagnosis of MDS with basophilia, but patient died within two months of diagnosis showing its poor prognosis. To increase awareness of the prognostic significance of MDS with basophilia, we report a case of MDS with basophilia which is a rare entity and showed poor prognosis.

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INTRODUCTION

Myelodysplastic syndromes (MDS) are a group of myeloid neoplasms characterized by abnormal differentiation and maturation of myeloid cells, reduced bone marrow (BM) function, and a genetic instability with enhanced risk to transform to secondary acute myeloid leukemia (AML) (Hofmann, 2005; Strom, 2008; Bennett, 2005; Valent *et al*., 2007). MDS with basophilia is a rare condition and has yet to be classified under the 2016 World Health Organization classification. However, reports have described its prognostic significance and also as an independent risk factor for evolution to leukemia. To predict the prognosis of patients with MDS, the Revised International Prognostic Scoring System (IPSS-R) (Revised International Prognostic Scoring System) is used. The IPSS-R is based on Cytogenetics, BM Blast %, Hemoglobin, Platelet count, absolute neutrophil count (ANC) but neither the IPSS-R nor the World Health Organization (WHO) takes into account the basophilia.Here we describe a case of MDS with basophilia which is rare presentation and showing poor prognosis.

Case report: A 60 years old male was admitted to the hospital with complains of generalised body weakness, easy fatiguability and breathlessness on mild exertion for one month. The patient was also a known case of diabetes on medication for 9 years. Clinical examinations were within normal limit without any organomegaly. Initial investigations showed features of pancytopenia with basophil 12% on complete blood count (CBC) with haemoglobin level of 8.6 g/dl, platelet count of 21 × 10^3/ µl and absolute neutrophil count of 360 /µl. Reticulocyte count was less than 0.5%. Random blood sugar was 126mg /dl which was within normal limit. Serum erythropoietin, total iron, TIBC and LDH were done which were within normal limit. Peripheral blood smear- On microscopic examination PBS showed features of pancytopenia with basophils 16%. Bone marrow aspiration (BMA) –BMA smears were diluted with blood and showed myeloid and erythroid series cells with features of dysmyelopoiesis and severe degree of dyserythropoiesis. Quadrinucleate, pentanucleate and gigantoblasts forms of erythroblasts were also noted. Occasional megakaryocyte seen. Imprint smears (IS) –IS were normo cellular as per cell density with adipocytosis and showed erythroid hyperplasia with significant dysplasia in granulocyte and erythroid cells along with presence of blasts (8%). Quadrinucleate normoblasts and gigantoblasts were noted. Bone marrow biopsy (BMB) -BMB showed normo cellular marrow spaces with brisk erythropoiesis and prominent granulopoiesis. Megakaryopoiesis was depressed. The paratrabecular cuff was

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widened with many foci of abnormal localisation of myeloid precursors (ALIP). Reticulin stain showed MF grade 0.

Clot section: 9normocellular particles with many erythroid cells. No megakaryocyte was seen

RESULT

Based on morphological examination, following differentials were included

- Myelodysplastic syndrome with excess blasts-1
- Acute myeloid leukemia with t(6;9)

Chromosomal analysis and FISH study were done. The patient's karyotype was normal and FISH study showed 7q deletion in 35% of cells. This excluded AML with t(6;9), hence a diagnosis of MDS-EB-1 with basophilia was made.

Table 1. IPSS-R risk score

Variables	Value	Score
Cytogenetics	Del(7q)	2
BM blast %	8%	2
Haemoglobin	8.6 g /dl	1
platelets	21 x10^9/L	1
ANC	0.36x10^9/L	0.5
	Total IPSS-R risk score	6.5

The IPSS-R risk score was calculated which was 6.5 with very high risk category and median survival of 0.8 years. The patient was on follow up but he died within 2 months of diagnosis due to myocardial infarction.

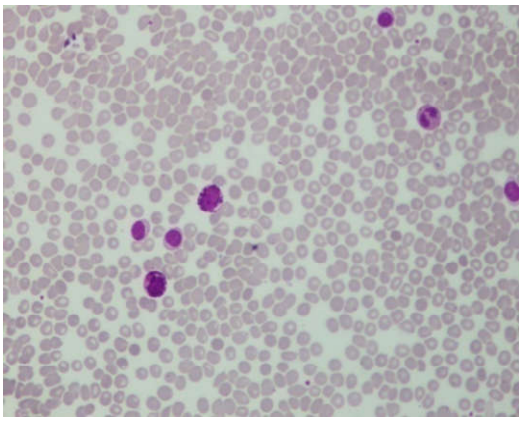

Figure 1. Peripheral blood smear showing basophilia

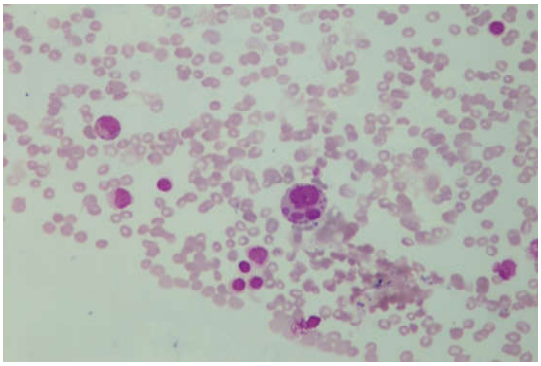

Figure 2. Bone Marrow Aspiration Smear showing quadrinucleate erythroblast

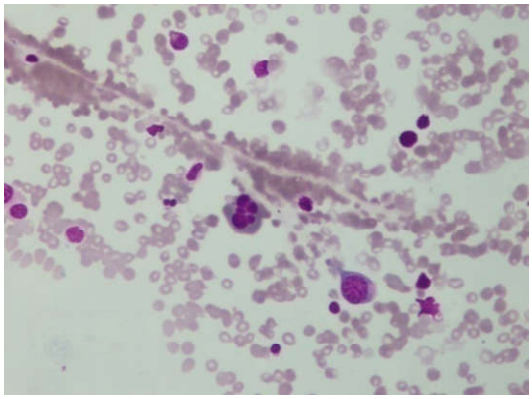

Figure 3. BMA Smear showing pentanucleate erythroblast

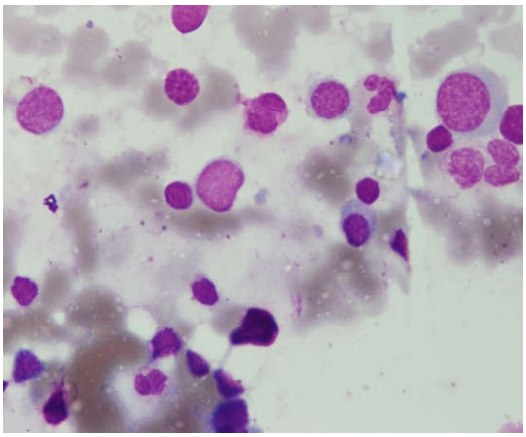

Figure 4. Imprint smear showing blasts

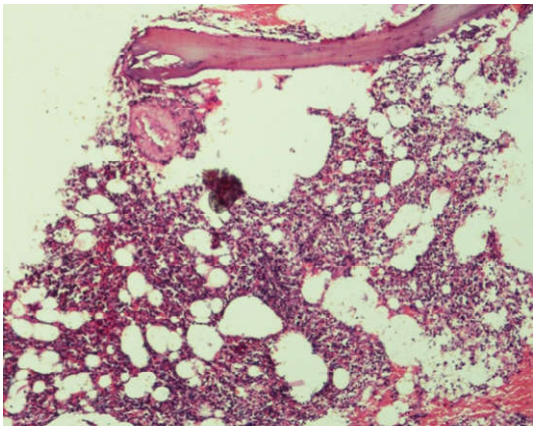

Figure 5. Bone Marrow Biopsy showing normocellular marrow

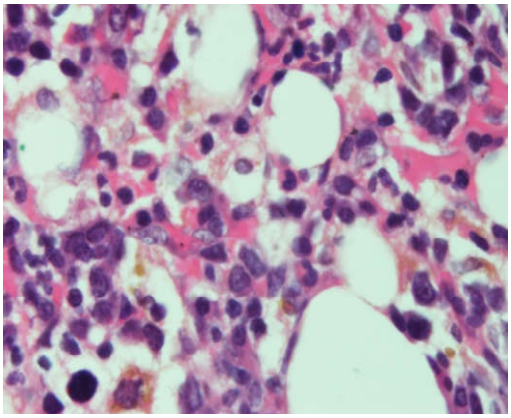

Figure 6. BMB showing ALIP

The IPSS-R risk score was calculated which was 6.5 with very high risk category and median survival of 0.8 years. The patient was on follow up but he died within 2 months of diagnosis due to myocardial infarction.

Figure 7. FISH study showing 7q deletion

DISCUSSION

- Myelodysplastic syndrome (MDS) is characterized by various forms of cytopenia with dysmorphic and ineffective hematopoiesis. This disease can affect patients of all ages but most commonly occurs in older patients with a mean age of 70 years (Yang, 2009; Bennett, 1982).
- In contrast, the data from Indian subcontinent shows that the median age at presentation is 45 years.[8]
- Although the prognostic value of neutropenia, thrombocytopenia and monocytosis have been documented, little is known about the impact of basophils. Morphologic assessment of BM and peripheral blood cells remains essential in the diagnosis and prognostication in MDS (Wimazal, 2010).

Friedrich *et al*. (2010) described the prognostic significance of eosinophilia and basophilia in MDS cases in their study. They found basophilia have poor prognostic significance. Takafumi *et al*[10] demonstrated that bone marrow basophilia was an independent risk factor for evolution to AML, bone marrow eosinophilia and basophilia in patients with MDS predict a poorer prognosis.

In our case based on morphology, cytogenetic study and FISH study a diagnosis of MDS-EB-1 was made and patient was referred to higher centre for further treatment but patient died 15 days after discharge due to myocardial in faction. The gold standard of risk assessment in MDS is the IPSS-R. In our case IPSS-R score was 6.5 with very high risk group category and median survival of 0.8 years.

Conclusion

MDS with basophilia is a rare condition which needs further studies about its prognostic significance. It also need to be evaluated as an independent risk factor for leukemic transformation. In our case MDS with basophilia showed poor prognosis.

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