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Myelodysplastic Syndrome/Myeloprolliferative Neoplasm Unclassifible

Dr P N Kadam* Dr Suvarnakar S V*, Drdeshpande S A**, Dr Panchal M***, Dr Hanmante R D***, Dr Shinde A P****, Dr Bhureapurva****
*(*Associate Professor, **Professor & Hod Pathology ,***Assistant Lecturer,**** Junior Resident.)*
Institution Dr.Shankarraochavangovt .Medical College, Nanded, India.

Abstract: MDS/MPN Unclassifiable is rare disease with mixed myelodysplastic and myeloproliferative features and cannot categorised into MDS OR MPN.WHO 2008 defined a MDS/MPN overlap category that includes
1) MDS/MPN Unclassifiable, 2)CMML, 3)JMML 4)Atypical CML,BCR-ABL 1 negative 5)Refractory anemia with ring sideroblasts and thrombocytosis MDS features with <20% blasts (not due to treatment)Prominent myeloproliferative features (leukocytosis,thrombocytosis,splenomegaly)No recent cytotoxic / growth factors Treatment Ph1negative,PDGFRA/PDGFRB negative, no del 5q, t(3;3) or inv 3 ORDenovo features of MDS/MPN that cant be assigned to another category

I. Introduction

MDS/MPN Unclassifiable is rare disease with mixed myelodysplastic and myeloproliferative features and cannot categorised into MDS OR MPN.

II. Objective

To review the clinical and pathological features of the disorder To discuss the prognosis and potential treatment approach.
MDS and MPN have overlapping features
WHO 2008 defined a MDS/MPN overlap category that includes:
MDS/MPN Unclassifiable
CMML
JMML
Atypical CML,BCR-ABL 1 negative Refractory anemia with ring sideroblasts and thrombocytosis

III. Case Report

50 yr serviceman vegetarian nonalcoholic came with C/o generalised weakness since 1 month, Ecchymoses ,petechiae since 15 days, swelling over right leg since 20 days,General Examination :- Pallor ++ Ecchymoses& petechiae on lower legs.Pulse :- 86/ min Systemic Examination :-CVS :- S 1 S 2 RS :-AEEBS PA :- moderate degree of hepatosplenomegaly On FNAC from swelling over right leg we found scattered large atypical cells (? blasts cells) with degenerated morphology that is suspicious of hematological malignancy. Patient further investigated

Investigation

1) CBC

Hb :– 7.4 gm%
TLC :- 130000 /cumm
DLC :- P-80% L-10% E-5% M-5%
Plat :- 57,000/cumm

2) Peripheral Smear

RBC :-Anisocytosis, Macrocytes, Microcytes,Moderate to severe degree of hypochromia
TLC :- Above normal

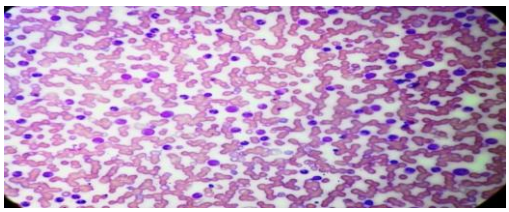


Figure :- 1 Blast cells and hypolobated myeloid cells in 40x

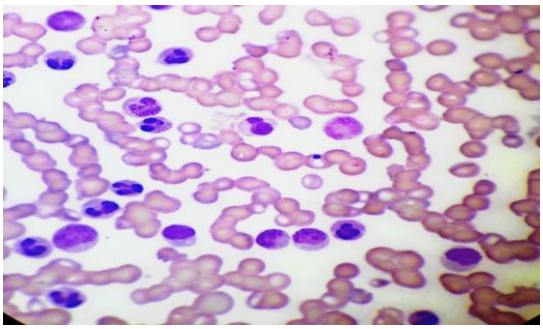


Fig 2 Hypolabulated and vacuolated Blast cells in oil immersion

DLC :- Blast like cells – 10 %, Myelocytes – 30 % ,Metamyelocytes -20 % , Band Forms – 5 % ,Mature polymorphs – 20 % Lymphocytes – 10 % ,Eosinophils – 5 %, Myeloid cells series shows hypolobulation with cytoplasmic vacuoles ,5-6 nucleated RBC /100 WBC Platelet severely depleted.

3) BM Aspiration

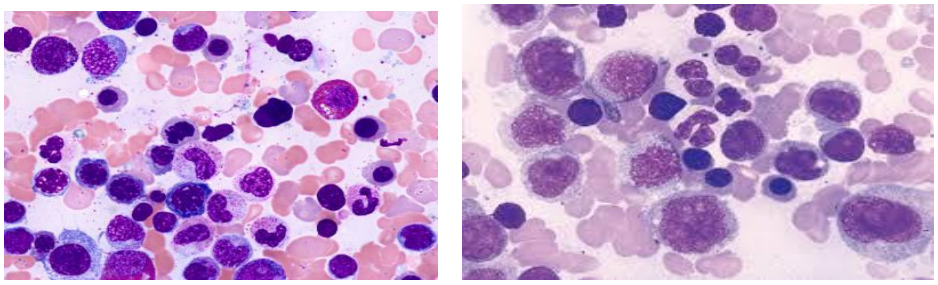


Fig 3 shows presence of blast cells ,erythroidprecurssor,eosinophilic blast cells.
Figure 4 :BM aspiration shows myeloblast and late erythroblastsBM Aspiration :-Shows hypercellular marrow with marked paucity of differentiation myeloid series noted. Erythroid series are decreased ,Megakaryocytes are not identified.Hypercellular marrow with myeloid preponderance & 11% blasts
Suspect MDS/MPN

4) BM Biopsy

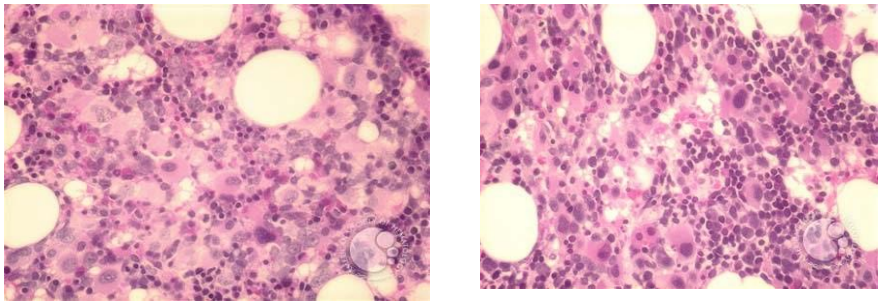


Figure :- 5 & 6 BM Biopsy Presence of blast cells hypolobulated myeloid series.
Consistent with AML

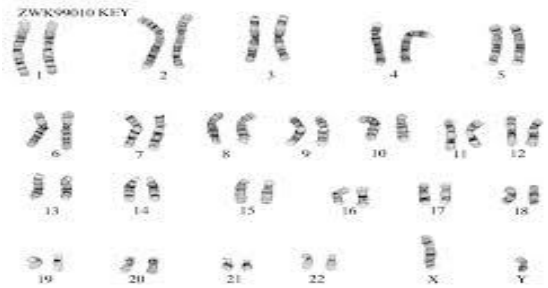
Hyperplastic bone marrow shows presence of Left shift of myeloid series,presence of pseudo pelgerhuet cells, blast cells. Erythroid series are decreased Megakaryocytes are not identified

5) Flow Cytometry

Morphological and immunophenotyping are consistent with MDS/MPN Unclassifiable,MPO +ve, with dim expression of CD 45 +ve CD 117 +ve compare to normal progenitors. CD 34 + ve,granulocytes are hypogranular in nature and show asynchronous maturation.
These findings are consistent with MDS-MPN

6) Molecular Cytogenetics (FISH)

No indication of deletion 5q (for RAEB,RARS,),7q (for JMML,CMML,RAEB,RARS) 20q, And Trisomy 8 (for CMML,MDS) ,along with no evidence of BCR/ABL fusion and negative for single round RT-PCR for b2a2 and b3a2 transcript.Also show no evidence of:t(8;21) (for AML M2),MLL translocation and inv (16) /t (16:16) for AML M4



Karyotyping suggestive of no chromosomal abnormality

IV. Discussion

Very rare disorder, 2% of cases in retrospective MDS series The cases classified as MDS/MPN U do not meet the criteria for CMML,JMML,atypical CML

Clinical presentation

Anemia ,+/- Macrocytosis,Proliferation in one of the myeloid lineages with dysplasia in at least 1 cell line.Thrombocytosis > 4.5lac/cmm, large and hypogranuloted orLeucocytosis ,WBC>13000/ cmm neutrophils may be dysplasticBlasts< 20%Cytochemical /immunophenotype findings similar to MPN and / or MDS Males =Females,60% symptomatic at diagnosis.Complication of cytopenias : fatigue ,bleeding,infection,Infiltration of spleen/liver,earlysatity Constitutional symptoms

V. Diagnostic Criteria

MDS features with <20% blasts (not due to treatment) Prominent myeloproliferative features (leukocytosis,thrombocytosis,splenomegaly)No recent cytotoxic / growth factors Treatment. Ph1negative,PDGFRA/PDGFRB negative, no del 5q, t(3;3) or inv 3 Or Denovo features of MDS/MPN that cant be assigned to another category

VI. Conclusion

MDS/MPN Unclassifiable clinically can present AML or CML morphologically immunophenotyping like FISH ,Cytogenetics are needed for confirmation of diagnosis and further management of patients

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