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Acute monoblastic leukemia as the rare presentation of AML with Myelodysplasia Related Changes (MRC)

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ABSTRACT

In 2008, the revised World Health Organization (WHO) classification of hematopoietic neoplasms introduced a new entity as a subcategory in the classification of acute myeloid leukemias (AML) entitled "acute leukemia with myelodysplasia-related changes" (AML-MRC) replacing the previously suggested entity in the 2001 classification "acute myeloid leukemia with multilineage dysplasia" (AML-MLD). Morphologic evaluation of dyspoiesis can be unreliable when there are very few residual hematopoietic cells and the blast count is high.Unlike the AML-MLD category, which relied mainly on morphology; this new AML-MRC category provides ample criteria for diagnosis, including cytogenetics. Acute myeloid leukemia with myelodysplasia related changes having unbalanced chromosomal abnormalities like 7q deletion are aggressive leukemias with a poor prognosis. We report one such rare case of acute myeloid leukemia presenting with extramedullary involvement and gingival swelling, morphologically and clinically resembling acute monoblastic leukemia but associated with 7q deletion.

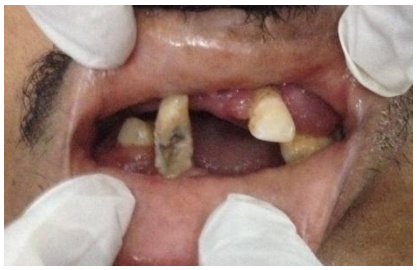
Key words: Acute myeloid leukemia, myelodysplasia, cytogenetics, 7q deletion.

INTRODUCTION

Leukemias are hematological disorders characterized by proliferation of abnormal hematopoietic stem cell. It can be of the myeloid or lymphoid origin. A significant percentage of acute leukemias do present with extramedullary involvement, commonest among them being acute myelomonocytic and monoblastic leukemias, more so with monoblastic leukemias.[1,2] Most commonly involved extramedullary sites are the liver, spleen, lymph nodes, mucosa, gingiva, skin etc. These leukemias can be associated with various chromosomal abnormalities.We report a case of acute myeloid leukemia presenting with extramedullary involvement associated with 7q deletion but morphologically and clinically resembling monoblastic leukemia. Such a case of acute myeloid leukemia with 7q deletion now falls into category of acute myeloid leukemia with myelodysplasia related changes according to the 2008 WHO classification.[3]

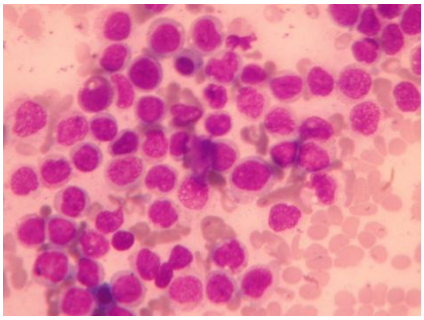
CASE REPORT

A 35 year old man, presented with three year history of swollen gums, loss of teeth and inguinal swelling (Figure 1). Figure 1: Clinical picture showing gum hypertrophy and loss of teeth.



He had been to dentist for his oral problems and was treated for gingivitis with no improvement. He was referred for our hospital for further management. On admission his blood counts were hemoglobin- 8.8gm/cu mm, total leucocyte count – 36,640 cells/cu mm and platelet-1.78 lakhs /cu mm.The peripheral smear showed predominance of monoblasts and promonocytes accounting for 85% of all nucleated cells (Figure 2).

Figure 2: Peripheral smear showing blasts with prominent multiple nucleoli and abundant cytoplasm, some of them also show cytoplasmic vacuoles.(May Grunwald Giemsa, X 100)



Bone marrow aspirate (BMA) and biopsy also revealed 80% monoblasts which were myeloperoxidase stain negative (Figure 3,4,5,6).

Figure 3: BMA shows sheets of blasts with suppression of erythroid and megakaryocytic series. (MGG , X40

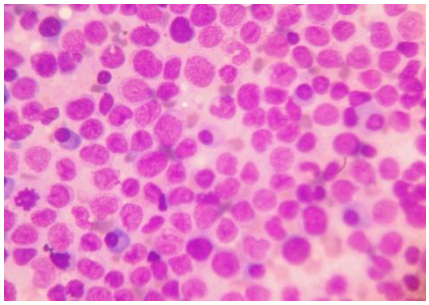


Figure 4: BMA shows monoblasts with multiple nucleoli abundant cytoplasm with scattered granules and vacuoles. (MGG,X100)

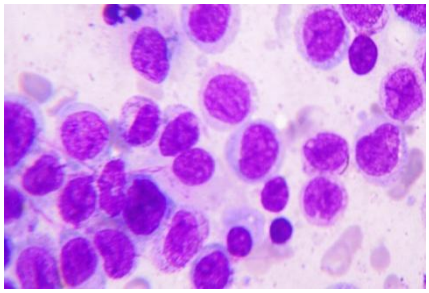


Figure 5: Bone marrow biopsy shows sheets of blasts. (H&E, X40)

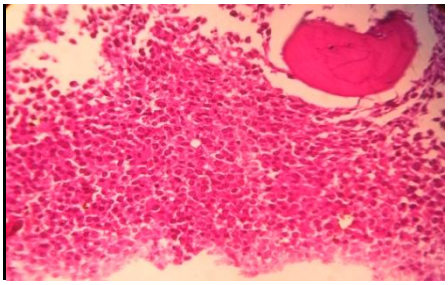
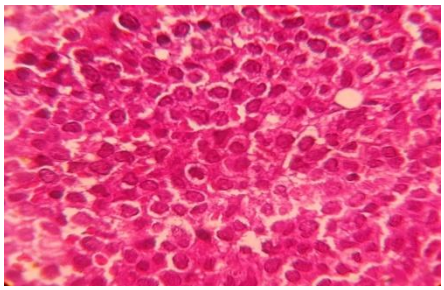
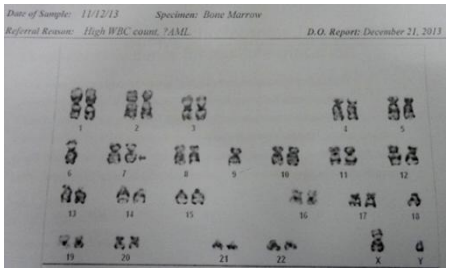


Figure 6: Bone marrow biopsy shows sheets of blasts. (H&E,X100)



A diagnosis of acute monoblastic leukemia was given based on the peripheral smear and bone marrow findings. Ultrasonography of abdomen revealed moderate splenomegaly. Flow cytometry done later revealed these bone marrow aspirate cells positive for CD33 and HLADR and negative for CD 13, CD 14 and CD 34 which is consistent with acute monoblastic leukemia. But cytogenetic study on bone marrow aspirate revealed deletion (7)(q22q32) in 5 out of 10 metaphases analysed (Figure-7).

Figure 7 : Cytogenetics report showing 7q(q22q32) deletion.



With all these investigations the case was diagnosed as AML with MDS associated cytogenetic abnormalities mainly due to 7q deletion. He was started on aggressive induction chemotherapy following which his gingival enlargement, nodular inguinal swelling and splenomegaly subsided.

DISCUSSION

For decades, morphology has been the cornerstone of diagnosis in AML. In 2008, WHO revised its classification of acute myeloid leukemia by paying attention to clinical relevance of cytogenetic abnormalities along with morphological, immunophenotypic and clinical information. Since then there has been a dramatic improvement in the cytogenetic and molecular understanding of AML. According to this new classification, acute myeloid leukemia with myelodysplasia related changes should satisfy atleast one of the following criteria: (1) AML with multilineage dysplasia - for any case to fall in to this category 50% of the cells in atleast two cell lines should have features of dysplasia like dysgranulopoiesis, dyserythropoiesis and dysmegakaryopoiesis.[4] The second criteria is (2) AML with MDS associated cytogenetic abnormalities - for any entity to be called AML with MDS associated cytogenetic abnormalities should have a complex karyotype or balanced translocations like t(11,16), t(3,21) or unbalanced translocations like 7q deletion,5q deletion, 13q deletion or 9q deletion.[4] The third criteria is (3)AML arising from a previously diagnosed case of MDS or MDS/MPN.[4]

Of these categories first and third, did not show much variation in patient survival compared to acute myeloid leukemia NOS (not otherwise specified),whereas second category showed a overall decrease in survival and effective disease free survival.[3] Usually these cases present either as acute myeloid leukemia undifferentiated or AML without maturation based on morphology. But in our case it was acute monoblastic leukemia, on both peripheral smear, bone marrow study and on flow cytometry too. Only when cytogenetics was

done that we were able to categorize this case under the entity of AML with myelodysplasia related changes which has a lower overall survival and effective disease free survival compared to the other categories. Therefore these patients warrant an alternative therapeutic approach like more aggressive chemotherapy or stem cell transplantation to improve their outcome.

CONCLUSION

Thus from our case it becomes obvious to do cytogenetic for every case of acute myeloid leukemia apart from the routine investigations like bone marrow study and flow cytometry where ever possible. This cytogenetic not only helps in definitive categorization but also aids to plan for an appropriate treatment protocol.

REFERENCES

1. Mark P, Ronald A, Damiano R, John Q, Valerie L, Sujata G. Acute myeloid Leukemia. With myelodysplasia related changes: A new entity defined by WHO 2008 classification of hemaopoietic and lymphoid neoplasms. Lab medicine.2009 ;40: 397-400
2. Gallipoli.P, Leach.M. Gingival infiltration in acute monoblasticleukaemia.BritishDental Journal. 2007; 203: 507- 09.
3. Heba N. Abdelrazik, Hala M, Farawila, Mai A Sherif, Mervat Al Ansary. Molecular characterization of chromosome 7 in AML and MDS patients. African Journal of Health Sciences.2006;13:33-42.
4. DA Arber, RD Brunning, A Orazi, BJ Bain. Acute myeloid leukemia with myelodysplasia related changes. In: Swerdlow S H, Campo E, Harris NL, Jaffe ES, Stein H, Thiele J et al, editors. WHO Classification of Tumors of Haematopoietic and Lymphoid Tissues. IARC: Lyon;2008.p.124 26.
5. Miriam Miesner, Claudia

Haferlach, Ulrike Bacher, Tamara Weiss, Katja Macijewski, Alexander Kohlmann et al .Multilineage dysplasia in acute myeloid leukemia correlates with MDS-related cytogenetic abnormalities and a prior history of MDS or MDS/MPN but has no independent prognostic relevance: a comparison of 408 cases classified as “AML not otherwise specified” or “AML with myelodysplasia-related changes”. Blood. 2010;116:2742-51.