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Case Report

Acute Promyelocytic Leukemia in a child with Congenital Baldness: A Novel Presentation

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Abstract: Acute leukemia is most common in adults with a worldwide incidence of 351,965 cases in 2012 with a male: female ratio of 1.4. We present the case of a 12 years old female child with congenital baldness who was admitted to our hospital with complaints of epistaxis, gum bleed and petechial rash. She was diagnosed as a case of acute promyelocytic leukemia(APL) on bone marrow biopsy. This is a rare finding not previously reported in literature.
Keywords: Acute Promyelocytic Leukemia, Congenital Baldness

Introduction

Leukemia is a group of malignant diseases involving the hemopoietic system and causing increased production of either the immature cells or the differentiated cells causing acute or chronic leukemia respectively. The diagnosis of leukemias involves a stepwise approach starting from the blood picture with peripheral film after the clinical suspicion of the disease. Chronic leukemias are usually diagnosed incidentally. Acute leukemia is the most common form of cancer in children accounting for almost 30 percent of childhood malignancies with acute lymphoblastic leukemia(ALL) being 5 times more prevalent than acute myeloid leukemia.[1]

Acute myeloid leukemia(AML) is most common in adults(2,3). In contrast to this high incidence in adults, AML constitutes less than 10 percent of acute leukemias in children less than 10 years of age.(3). More than 50 percent of the adults with acute myeloid leukemia carry chromosomal abnormalities like t(8;21)(q22;q22), t(15;17),t(8;21) or t(16;16).(4)Common clinical presentation is due to the replacement of bone marrow by blast cells, causing cytopenias, leading to the presenting features of fatigue, shortness of breath, infections and bleeding.[2,3]

The French-American-British(FAB) classification subtypes AML based on morphology and cytochemical staining with immunophenotypic data in some cases.

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Types M0 to M3 are granulocytic and are different on the basis of maturity of the cell type.M4 is both granulocytic and monocytic with at least 20% monocytic cells whereas M5 is predominantly monocytic. M6 shows erythrocytic differentiation with dysplastic features including megaloblastic change.M7 is megakaryocytic leukemia.[3]

The World Health Organization (WHO) classification differs from the FAB classification in its use of genetic analysis along with the morphological features.AML is classified into groups including AML with recurrent genetic abnormalities; AML with myelodysplasia related changes; Therapy related myeloid neoplasms; AML not otherwise specified; Myeloid sarcoma and Myeloid proliferations related to Down's syndrome. The most commonly identified genetic abnormalities in AML are t(8;21) (q22;q22), inv(16) (p13.1q22) or t(16;16) 9p13.1;q22), t(15;17) (q22;q12) and t(9;11) (p22;q23). These lead to the development of leukemia in the affected individual.[4] The translocation in APL is t(15;17) and it presents with disseminated intravascular coagulation.

Alopecia is 'the loss of hair' from part of the body, usually from the head. It is mostly acquired; secondary to malnutrition, hypothyroidism, chemotherapy or certain fungal infections. Cases have been reported of few neonates with alopecia but none having association with APL have been seen

Case Report

Our patient was a 12 years old female, resident of Chakwal presented with the complaints of

1. Bleeding from nose and mouth for 1 day;
2. Rash all over the body for 1 day;

3. Fever off and on for past 2 weeks along with cough and it was undocumented at the time of admission. Her bone marrow biopsy was done on 8th April, 2017.

Her past history was non-significant. One unusual physiological finding in our patient was her congenital baldness. She did not have any eyebrows or eyelashes. According to the mother there were other kids in the area without hair since birth.

Her family history was negative for any blood disorder. She was transfused with a total of 4 Red cell concentrates and 14 platelet units. Last red cell transfusion was done on 1st April, 2017. For her treatment during her admission she was being given antibiotics, Vitamin K and Hydrocortisone Sod Succinate (Solu-Cortef) along with nebulization.

Examination showed a young female child lying comfortably on the bed with no visible signs of respiratory distress. She was pale with no hair on the visible parts of her body though according to the mother she had a normal I.Q. level and was a student. Her systemic examination was unremarkable. There were no lymph nodes or hepatosplenomegaly. Bone marrow biopsy was requested with the suspicion of Immune thrombocytopenic purpura(ITP).

Her laboratory investigations available at the time of biopsy were;

LABORATORY TEST	REPORT
Complete Blood Picture	**Total leucocyte count** 8.1x10^3/µL **Hemoglobin** 8.1g/dL **Platelets** 10x10^3/µL **Mean corpuscular volume** 92.8 fL **Mean corpuscular hemoglobin** 30.6 pg **Mean corpuscular hemoglobin concentration** 32.9g/dL
Reticulocyte count	3%(0.5-1%)
Renal Function Tests	Normal
Liver Function Tests -Serum Total Bilirubin -Alanine Aminotransferase -Alkaline Phosphatase	 1.9mg/dL(Normal= upto 1 mg/dL) Normal Normal
Random Blood Sugar	Normal
Serum Electrolytes	Normal
Serum Calcium	0.92mmol/L (Normal=1-2.4mmol/L)
Prothrombin Time	18 seconds(Normal Control 13 seconds)
Activated Partial Thromboplastin Time	42 Seconds(Normal Control 32 seconds)
Serum Albumin	2.5g/dL(Normal3.5-5g/dl)
Serum vitamin B12	175pg/mL(Normal=189-883pg/mL)
Serum Folic acid	9.2ng/mL(Normal)
Blood Culture	Yielded No Growth

Bone marrow biopsy was performed from the posterior superior iliac spine which showed abnormal promyelocytes and blasts. The blasts gave positive Sudan Black staining (more than 3%) and negative PAS (periodic acid Schiff) with Auer rods in few blasts and a final diagnosis of APL was made.

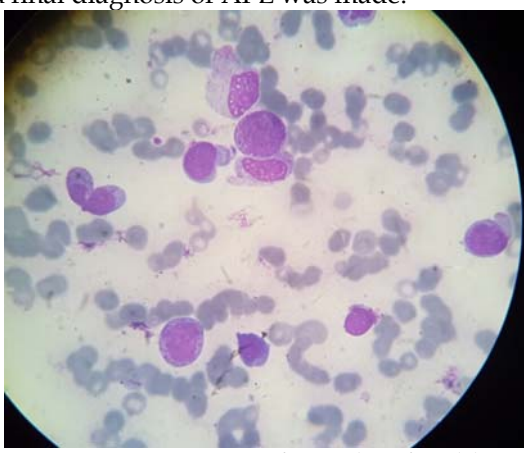

Figure-1: Bone Marrow aspirate showing blasts and promyelocytes (X 1000)

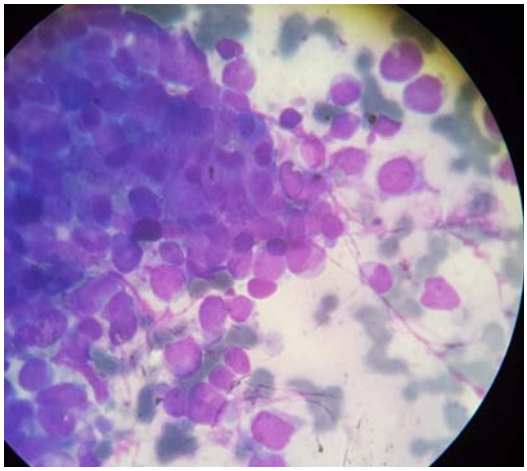

Figure-2: Hemophagocytosis (X 1000)

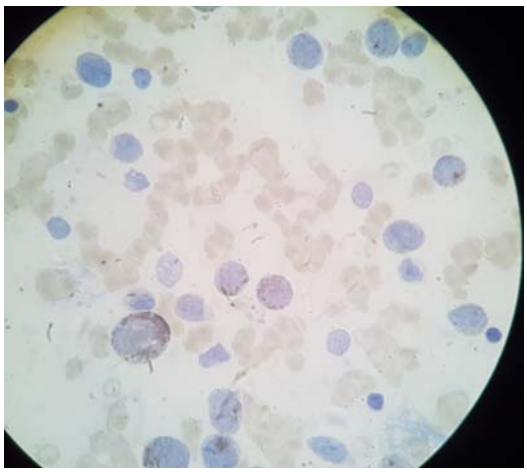

Figure 3: Positive Sudan Black staining (X 1000)

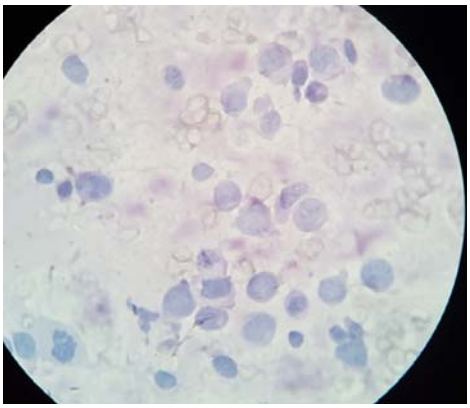

Figure 4: Negative PAS staining (X 1000)

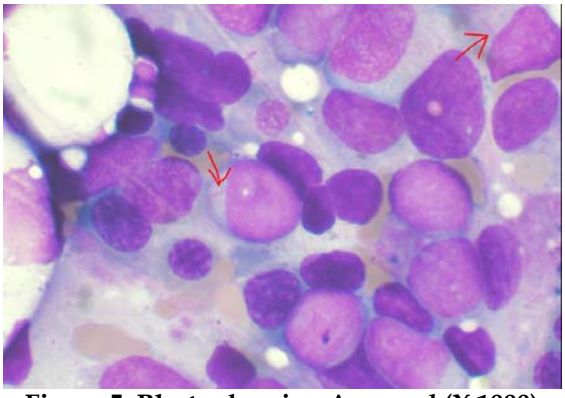

Figure 5: Blasts showing Auer rod (X 1000)

Discussion

Acute leukemia is the most common form of cancer in children almost 30 percent of childhood malignancies with acute lymphoblastic leukemia(ALL) being 5 times more prevalent than acute myeloid leukemia.[4-7]

Very few cases of AML at this young age are reported; ALL being the commoner childhood leukemia.[6,7] In pregnant women with promyelocytic leukemia maternal and fetal out comes is better if diagnosis is made early and immediate treatment is instituted.[8]

Another 53 years old man presented with fatigue and dyspnea and was diagnosed as a case of AML. Presenting features are due to decreased particular cell lines and include fatigue due to anemia and bruises or increased tendency to bleed due to decreased platelets. Our patient unfortunately expired in the I.C.U. of the hospital and genetic analysis could not be done to find out the particular mutation. According to the mother of the child there were other children in the area with similar congenital absence of hair. This needs to be investigated in relation to similar ill health to see if it has some relation with AML occurrence and prevalence in the area.

Conclusion

Diagnostic and treatment facilities should be available at a tertiary care hospital like ours where people are referred from periphery for diagnosis and treatment. Unfortunately, at present we don't have DNA analysis techniques available at PIMS neither do we have treatment facilities for AML available at our hospital so the patients are referred to other tertiary care or specialty dealing hospitals.

References

1. Dorantes-acosta E, Arreguin-gonzalez F, Rodriguez-CA, Sadowinski S, Pelayo R, Medina-sanson A. Acute myelogenous leukemia switch lineage upon relapse to acute lymphoblastic leukemia : a case report. 2009;5:1–5.
2. Sartor C, Papayannidis C, Abbenante MC, Curti A, Polverelli N, Ottaviani E, et al. A case report of acute myeloid leukemia and neurofibromatosis 1 on co m m er ci al us e on l on er al. 2013;5:1–2.
3. Report AC, Myeloid A, Fab L. Case Report. 2014;4:2–4.
4. Li L, Zhou F, Hou J. Case Report chromosome translocation: a case report and literature review. 2012;2(4):265–8.
5. Kumar H, Buch A, Iqbal B, Kakrani AL. Hypoplastic acute myeloid leukemia-M4 : A rare case report. 2016;9(1):2016–8.
6. Jazilah W, Ariffin W, Frcp E. M3-Variant of Acute Promyelocytic Leukaemia A Case Report in a Malay Boy. 1995;50(1):105–7.
7. Anirudha V. Kushtagi , Mihir J. Bhalodia , Mahesh H. Karigoudar , Jyotirling Savle. Acute Promyelocytic Leukemia in 4 year old female Child - A Case Report.JKIMSU July-Dec 2014;3(2):140-43.
8. Sanz MA, Montesinos P, Casale MF, Díaz-Mediavilla J, Jiménez S, Fernández I, Fernández P, González-Campos J, González JD, Herrera P, de Lisa E, Olave T, Rojas R, Salamero O, Sayas MJ, Pellicer A, Perales A. Maternal and fetal outcomes in pregnant women with acute promyelocytic leukemia.Ann Hematol. 2015 Aug;94(8):1357-61. doi: 10.1007/s00277-015-2372-5. Epub 2015 Apr 25.PMID:25911134

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