THE MYELODYSPLASTIC SYNDROME AND PRELEUKEMIA

ESSAY
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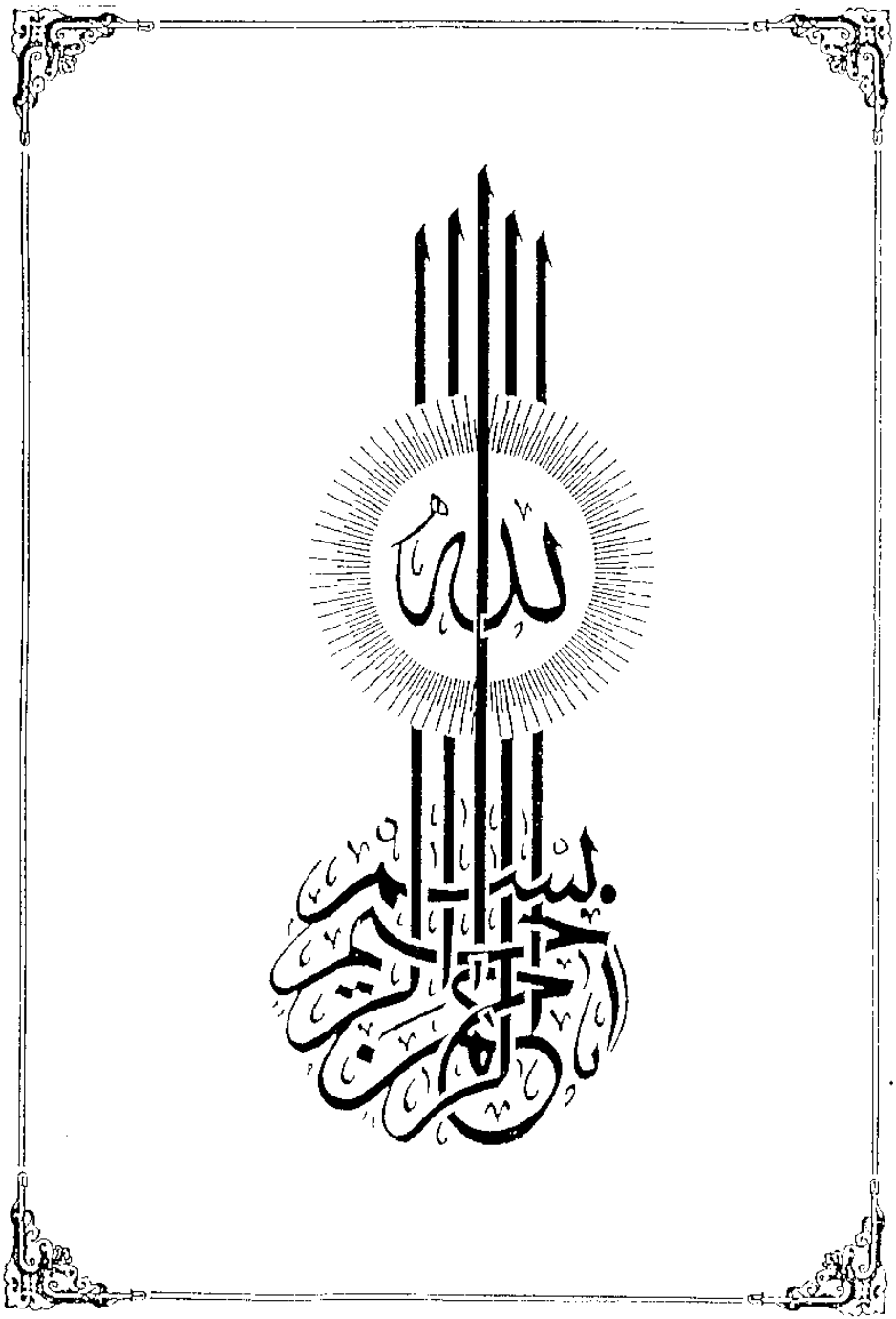
بسم الله الرحمن الرحيم

قَالُوا سُبْحَانَكَ لَا عِلْمَ لَنَا
إِلَّا مَا عَلَّمْتَنَا إِنَّكَ أَنْتَ
الْعَلِيمُ الْحَكِيمُ.

صدق الله العظيم

سورة البقرة آية ٣٢



TO MY FAMILY

WITH ALL MY LOVE

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Anan Beecher

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LIST OF ABBREVIATIONS

AIDS : Acquired immune deficiency syndrome.

AISA : Acquired idiopathic sideroblastic anaemia.

AL : Acute leukemia.

ALIP : Abnormal localization of immature precursors.

AML : Acute myeloid leukemia.

ANLL : Acute non lymphocytic leukemia.

bcr : break point cluster region.

BFU-E : Burst forming unit - Erythroid.

BM : Bone marrow.

CD : Cluster of differentiation.

CFU-E : Colony forming unit - Erythroid.

CFU-GEMM : Colony forming unit - Granulocytic, Erythroid, Monocytic, Macrophage.

CFU-GM : Colony forming unit - Granulocyte monocyte.

CFU-Meg : Colony forming unit - Megakaryocyte.

chr : Chromosome.

CMML : Chronic myelomonocytic leukemia.

CML : Chronic myeloid leukemia.

CSA : Colony stimulating activity.

CSF : Colony stimulating factor.

DNA : Desoxyribonucleic acid.

FAB : French - American - British Cooparative Group.

GM-CFC : Granulocyte/ macrophage - colony forming cells.

GM-CSF	:	Granulocyte monocyte colony stimulating factor.
HbF	:	Faetal hemoglobin.
inv	:	inversion.
I.V.	:	Intravenously.
LDAC	:	low dose cytosine arabinoside.
M-CSF	:	macrophage colony stimulating factor.
MCV	:	Mean cell volume.
MDS	:	Myelodysplastic syndrome.
2°MDS/AL	:	Secondary MDS/AL.
MOPP	:	Nitrogen Mustard - Oncovin - Prednisone and Procarbazine.
N/C	:	Nuclear/ cytoplasmic.
NHLs	:	Non Hodgkin lymphomas.
P	:	Short arm of chromosome.
P.B.	:	Peripheral blood.
ph'	:	philadelphia chromosome.
PK	:	Pyruvate kinase.
PL	:	Preleukemia.
PL/L	:	Preleukemia/ leukemia.
PLS	:	Preleukemic syndrome.
5q-	:	Deletion of long arm of chromosome 5.
RA	:	Refractory anaemia.
RAS	:	Refractory anaemia with ring sideroblasts.
RAEB	:	Refractory anaemia with excess blasts.

RAEBt : Refractory anaemia with excess blasts in transformation

RNA : Ribonucleic acid.

SLE : Systemic lupus erythematosis.

t : Translocation.

WBC : White blood cells.

CONTENTS

INTRODUCTION AND AIM OF THE WORK

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Over the past decade, hematologists have become increasingly aware of patients whose have had refractory cytopenia accompanied by dysplasia of one or more of their hemopoietic cell lineages *(Oscier, 87)*.

Although this early observation was confirmed by a number of workers, the concept of preleukemic state was only established in 1953 by Block and Co-workers.

A set of diagnostic criteria of the preleukemic syndrome or hemopoietic dysplasia were established to identify prospectively those patients with a very high likelihood of developing acute myeloid leukemia *(Brennan and Lichtman, 1990)*.

In 1982, the FAB group *(Bennett et al., 1982)* introduced proposals for the classification of the myelodysplastic syndrome (MDS) and defined five groups:

1- Refractory anaemia (RA).

2- Refractory anaemia with ring sideroblasts (RAS).

3- Refractory anaemia with excess blasts (RAEB).

4- Refractory anaemia with excess blasts in transformation (RAEBt).

5- Chronic myelomonocytic leukemia (CMML).

The aim of this study is to identify precisely myelodysplastic syndrome as regards: Classifications, pathogenesis, clinical variaties, heamatological findings, cytogenetic study, prognosis and treatment.

REVIEW OF LITERATURE

MYELODYSPLASTIC SYNDROME AND PRELEUKEMIA

DEFINITION:

Myelodysplastic syndrome (MDS) and preleukemic syndrome (PLS) are synonymous for a group of clonal proliferative disorders of the bone marrow that are characterized by dysmyelopoiesis and peripheral blood cytopenias. These disorders may progress to overt acute non lymphocytic leukemia (ANLL) or cause death of the patient secondary to complication of ineffective hematopoiesis (*Pierre, 1986*).

HISTORY AND NOMENCLATURE:

Patients who would now be recognized as having MDS have been described since the turn of this century, but evaluation of early reports is hindered by the fact that different haematologists have emphasized particular aspects of the condition and a confusing nomenclature has arisen, with some terms being synonymous and others reflecting different phases in the natural history of MDS (*Oscier, 1987*).

In the 1930s there was much interest in patients with anaemias which were refractory to haematinics such as iron, vitamin B_{12} and folic acid.. *Rhoads and Barker (1938)*

described 100 cases of refractory anaemia some were associated with and secondary to other diseases, while others were thought to be primary haematological disorders. this anaemia was often associated with leucopenia and thrombocytopenia.

Chevallier - in 1942 discussed the "odo-leukemias". He chose the Greek word odo, meaning threshold, to highlight disorders that are on the threshold of leukemia.

Hamilton-Paterson (1949) described three patients who presented with refractory anaemia and subsequently developed acute myeloid leukemia. Seven years later *Bjorkman (1956)* delineated a subgroup of refractory anaemia characterized by the presence of ring sideroblasts in the marrow, and one of his four cases evolved into acute myeloid leukemia (AML).

In the 1950s and early 60s there was an increasing awarness of elderly patients with a modest increase in blast cells in the marrow in association with a peripheral cytopenia, whose illness pursued a chronic course before terminating in acute leukemia (AL). This condition was variously described as preleukemic acute human leukemia (*Block et al., 1953*), low percentage leukemia (*Dameshek & Gunz, 1958*) and smouldering acute leukemia (*Rheingold et al., 1963*).