26.4±2.7 mg/m^2/day. ATRA syndrome was observed in 10/34 (29.4%) and was non-fatal. Prophylactic dexamethasone was used in 14 (73.7%) out of 19 high-risk patients. Sixteen (39%) patients died; of which 15 (94%) died within 15-days of diagnosis. The cause of death included, intracranial bleed (82%) and septicemia (18%). There was merely one mortality beyond induction from febrile neutropenia following first cycle of consolidation. The risk category did not predict mortality (p=0.52). There was no correlation of <15 day mortality with platelet count (p=0.61) or coagulopathy (p=0.12). No patient died or abandoned treatment following 2-months of treatment. The median follow-up of patients who continued treatment beyond 2 months was 22 months (range: 1.5-141). No patient relapsed. The 10-year OS as well as EFS of the cohort (n=41) was 58.4±8% and for patients who survived ≥15 days from diagnosis (n=26) OS/EFS was 95.2±4.6% (p=0.01).

Conclusion: The EFS of all children with APML, and of those who survived ≥15 days from diagnosis was 58.4±8% and 95.2±4.6%, respectively (figure 1). Early (<15 days from diagnosis) mortality is predominantly caused by intracranial hemorrhage and is a major impediment to survival in children with APML.

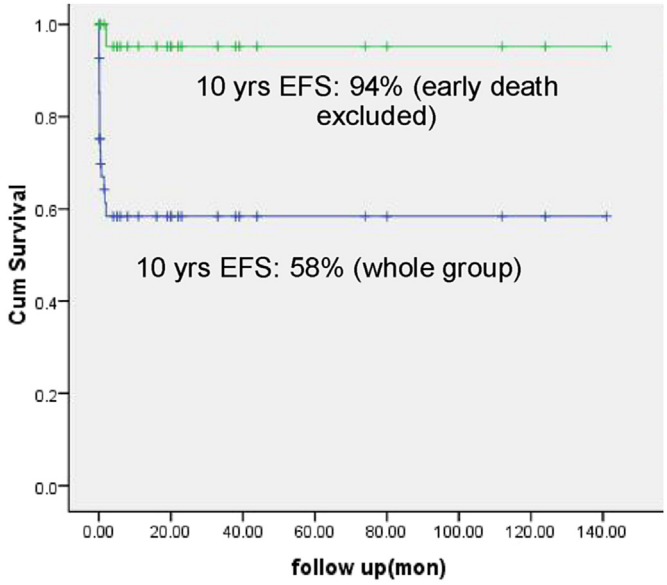

Figure 1. Comparison of survival between entire cohort and those excluding early mortality.

MM-1_V1.5
INCIDENCE, RISK FACTORS AND OUTCOME OF THERAPY RELATED ACUTE MYELOID LEUKAEMIA: SINGLE CENTRE EXPERIENCE FROM A PAEDIATRIC ONCOLOGY UNIT

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Abstract

Introduction: Therapy related acute myeloid leukaemia (t-AML) is a rare, though devastating late effect of cancer treatment. Alkylating agents, DNA topoisomerase II inhibitors and radiotherapy have been mainly implicated as causative agents. Usually these are refractory to antileukemic therapy and warrants allogenic bone marrow transplant.

Material and Methods: This is a retrospective analysis of patients less than 18 years of age treated in paediatric Hematology-Oncolgy unit at our institute between January 1996 and December 2015. Amongst these, patients who developed t-AML were identified. Information pertaining to primary malignancy and its treatment along with clinical, hematologic, cytogenetic features and outcome of t- AML were collected.

Results: Amongst 1543 children treated during this period, eight patients developed t-AML (6 males and 2 females) with a median age of 15.5 years (range, 4-22 years). Overall incidence of t-AML was 0.5% [0.7% (4/515) in solid tumours compared to 0.4% (4/1028) in leukemia/lymphoma, P=0.45] with a median follow up of 7 years. Primary malignancy included sarcoma in 4 patients (one each with osteosarcoma, rhabdomyosarcoma Ewings and synovial sarcoma) and hematolymphoid malignancy in 4 patients [two each with B-non-Hodgkin lymphoma (NHL) and acute lymphoblastic leukemia (ALL)]. Presenting features of t-AML were non resolving fever (6/8), bone pains (3/8), gingival hyperplasia (5/8) and bleeding manifestation (4/8) with a median latency period of 24 months (range 16.5-62 months). Only 1/8 patients had a preleukemic phase. The FAB morphology was M2 (1/8) and M4/M5 (7/8). Of 6 patients whom cytogenetic studies were available, 4 had MLL rearrangement. Total cumulative cyclophosphamide equivalent dose was 28.3 g/m^2, 27.7 g/m^2 and 13.1g/m^2 in patients with Ewings sarcoma, rhabdomyosarcoma and synovial sarcoma respectively and 3 g/m^2, 6.8 g/m^2 in patients with ALL and B-NHL (group C). The cumulative dose of etoposide was 4 g/m^2 in patients with Ewings sarcoma and rhabdomyosarcoma, 1g/m^2 in synovial sarcoma and 2.5 g/m^2 in B-NHL (group C). The median cumulative dose of anthracyclin was 367mg/m^2 (range, 300- 375mg/m^2) in sarcoma patients and 240mg/m^2 (range, 180-240mg/m^2) in leukemia/lymohoma patients. Two patients received radiotherapy to primary site [1 RMS (50Gy), 1 Synovial sarcoma (45Gy)]. Treatment for t-AML was opted by 5/8 patients out of which three achieved complete remission and one is currently on induction. Three patients died [2 toxic death (1 during induction, 1 during remission) and 1 relapse/progressive disease]. One patient (post allogenic BMT) is alive and disease free at 25 months of follow up.

Conclusions: We observed a high incidence of t-AML in children with pediatric sarcoma compared to haematological malignancies. Shorter latency period, absence of preleukemic phase, presence of MLL gene implicates epipodophyllotoxin to be the probable causative agent. In view of poor outcome with conventional therapy, novel strategies need to be considered.

Keywords: t-AML, children, leukemia, solid tumor, alkylating agents

MM-1_V1.6
IS COEXISTING CYTOMEGALOVIRUS INFECTION AND JUVENILE MYELOMONOCYTIC LEUKEMIA ASSOCIATED WITH POOR PROGNOSTIC OUTCOME?

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Abstract

Background: Juvenile myelomonocyticleukemia (JMML) is a potentially fatal, rare clonal haematopoietic myeloprolifertive/ myelodysplastic group of disorders of childhood characterized by excessive proliferation of the granulocytic and monocytic lineages. JMML is known to mimic/co-exist with viral infections which further makes the diagnosis difficult and delayed. The aim of this study is to highlight the high incidence of JMML coexisting with viral infections, in particular herpes group of viruses in Indian scenario leading to diagnostic dilemma and subsequent delay in management.

Material and methods: Retrospective analysis of case records of eleven patients diagnosed with was done. The diagnosis was established based on International JMML working group diagnostic criteria.

Results: JMML constituted 3% of all haematological malignancies and 1% of all childhood malignancies reporting to our centre. Three of the eleven children with JMML presented with co-existing Cytomegalo virus infection while two had co-existing EBV infection. All three patients of JMML with co-existing CMV infection died early due to disease progression while those with co-existing EBV and EBV and parvo virus B19 infection are alive after 24 months and 8 months of disease presentation.

Conclusion: JMML with co-existing CMV infections leads to poor outcome and it must be considered as a differential diagnosis in children as it mimics /co-exists with viral infection.

Our study is first case series on JMML with coexisting viral infections from Indian subcontinent.