

ABSTRACT

Background

Acute promyelocytic leukaemia (APL) is a subtype of acute myeloid leukaemia (AML). APL is a medical emergency due to its high early mortality rate, most commonly due to bleeding complications resulting from an associated coagulopathy including a DIC (disseminated intravascular coagulation). However, APL occurs predominantly in young patients and is eminently treatable with supportive care, all-trans retinoic acid (ATRA) and appropriate chemotherapy.

Aims and Objectives

- To investigate the demographic and clinical profile of patients diagnosed with APL during the period 01/01/1994 to 31/12/2019.
- To determine the factors responsible for morbidity and mortality.
- To investigate the relationship with HIV (human immunodeficiency virus).
- To describe the management and outcome of the patients, including response to treatment and survival.

Patients and Methods

This was a retrospective study in which the records of patients diagnosed with APL from 01/01/1994 to 31/12/2019 at the Clinical Haematology Unit, Department of Medicine, Chris Hani Baragwanath Academic Hospital were assessed. Data relating to patient demographics, clinical presentation, laboratory parameters, management and outcomes was reviewed and analysed.

Results

There were 79 evaluable patients included in this study. Although 16.87% of the patients with APL were HIV seropositive, there was no clear association with HIV, implying that the relationship between the two diseases is likely coincidental. The APL coagulopathy was evident at presentation, with 82.05% of patients presenting with bleeding, and 4.29% with thrombosis. Laboratory evaluation revealed that 93.42% of patients had anaemia and 97.37% had a

thrombocytopenia. 63.93% of patients had evidence of a DIC. There was rapid improvement of all of these parameters following initiation of therapy. The Sanz risk score was used to assess risk, and 26.32% of patients had a high-risk APL. There was also a high percentage of patients with an immunophenotype associated with a high white cell count (WCC) and a poorer prognosis.

Management

Management involved supportive therapy with blood and blood products, and specific therapy, most commonly with ATRA and daunorubicin during induction and consolidation. Maintenance therapy included ATRA, methotrexate and mercaptopurine. There were 21 patients who received re-induction therapy, five of whom were for failed induction and the remaining patients for relapsed disease. Early complications, defined as complications occurring within the first 30 days from initiation of therapy, were found in 89.33% of patients, and 83.54% of these were bleeding complications. Thirty-seven patients achieved remission following induction therapy. At the last date seen, 39.24% of patients were in remission, 49.37% of patients had died and 3.8% were experiencing relapsed disease. Overall, the median survival was 7.8 months, which improved to 43.7 months once early mortality was excluded. There was no statistically significant difference in the survival time between HIV seropositive and seronegative patients. There was also no statistically significant difference in survival time across the low, intermediate and high-risk groups as calculated by the Sanz score.

Conclusion

Acute promyelocytic leukaemia (APL) most commonly presents with bleeding manifestations, anaemia, thrombocytopenia and DIC. These complications contribute to a high early mortality rate. Therefore, recognition and awareness of this entity, early diagnosis and timeous referral is of paramount importance in order to decrease the high early mortality of APL, a malignancy that is eminently treatable with appropriate supportive care and specific therapy.