Journal of the Egyptian National Cancer Institute (2014) **26**, 67–71

Cairo University

Journal of the Egyptian National Cancer Institute

www.nci.cu.adu.eg
www.sciencedirect.com

Full Length Article

A descriptive study of plasma cell dyscrasias in Egyptian population

Neemat M. Kassem [a], Hamdy EL Zawam [b], Heba A. Kassem [a,*], Tamer EL Nahas [b], Noha M. El Husseiny [d], Hamdy Abd El Azeeim [b,c]

[a] *Clinical Pathology Department, Kasr El-Aini Centre of Clinical Oncology & Nuclear Medicine (NEMROCK), Faculty of Medicine, Cairo University, Cairo, Egypt*
[b] *Clinical Oncology Department, Kasr El-Aini Centre of Clinical Oncology & Nuclear Medicine (NEMROCK), Faculty of Medicine, Cairo University, Cairo, Egypt*
[c] *Cairo Cure Center, Cairo, Egypt*
[d] *Clinical Hematology Department, Faculty of Medicine, Cairo University, Cairo, Egypt*

Received 1 July 2013; accepted 17 September 2013
Available online 17 December 2013

KEYWORDS

Plasma cell dyscasias;
Multiple myeloma;
Monoclonal gammopathy

Abstract *Background:* Plasma cell dyscrasias (PCDs) refer to a spectrum of disorders characterized by the monoclonal proliferation of lymphoplasmacytic cells in the bone marrow and, sometimes, tissue deposition of monoclonal immunoglobulins or their components. These disorders include multiple myeloma (MM) and Waldenström's macroglobulinemia, as well as rare conditions such as light-chain deposition disease (LCDD) and heavy-chain diseases (HCDs). The worldwide annual incidence of MM is estimated at 86,000, which is approximately 0.8% of all new cancer cases.

Purpose: Our retrospective study aims to highlight the immunologic and epidemiological features of PCDs mainly MM in Egyptian patients and compare our results with those of other populations.

Methods: Two hundred seventeen Egyptian patients with PCD were enrolled in the study. Serum, urine protein electrophoresis and immunofixation were used to demonstrate M protein.

Results: One hundred thirty-eight patients (63.6%) had IgG monoclonal band, 38 patients (17.5%) had IgA, 12 patients (5.5%) had Waldenström's macroglobulinemia (IgM monoclonal band) and 29 patients (13.4%) were light chain myeloma. One hundred fifty-one (70%) were Kappa chain positive and 66 patients (30%) were lumbda positive. Conventional cytogenetics was available for 40 patients; of them12 patients (30%) showed 13q-. Mean OS was 37.5 months (1–84 months). Survival analysis was statistically insignificant according to age, sex and ISS or type of treatment (P value > 0.05).

* Corresponding author. Tel.: +20 2 01144829798.
E-mail address: heba.kasem@hotmail.com (H.A. Kassem).

Peer review under responsibility of The National Cancer Institute, Cairo University.

Production and hosting by Elsevier

 http://dx.doi.org/10.1016/j.jnci.2013.09.002

Conclusion: Long term follow up is required to further define the role of different therapeutic lines of treatment including ASCT in the various stages of PCD based on OS data.

Introduction

Plasma cell dyscrasias (PCDs) constitute a broad spectrum of diseases characterized by clonal proliferation and accumulation of cells producing monoclonal immunoglobulins (M component) and include monoclonal gammopathy of undetermined significance (MGUS), multiple myeloma (MM), smoldering multiple myeloma (SMM), plasma cell leukemia (PCL), Waldenström's macroglobulinemia (WM), POEMS syndrome, plasmacytoma, heavy chain disease (HCD), and amyloidosis [1].

Multiple myeloma is the most serious and prevalent plasma cell dyscrasia and accounts for approximately 10% of all hematologic cancers [2,3]. It usually evolves from an asymptomatic premalignant stage of clonal plasma cell proliferation termed "monoclonal gammopathy of undetermined significance" (MGUS). MGUS is present in more than 3% of the population above the age of 50 and progresses to myeloma or related malignancy at a rate of 1% per year [4,5]. In some patients, an intermediate asymptomatic but more advanced premalignant stage, referred to as "smoldering multiple myeloma", is clinically recognized [6].

The worldwide annual incidence of MM is estimated at 86,000, which is approximately 0.8% of all new cancer cases [7]. Approximately 63,000 deaths are reported annually, which is 0.9% of all cancer-related deaths [7]. The American Cancer Society estimates that in 2013, there will be 22,350 cases of MM diagnosed (12,440 in men and 9910 in women) and 10,710 deaths related to MM (6070 in men and 4640 in women) in the United States. The incidence rate of multiple myeloma was significantly higher among people living in urban areas than those from rural areas. Residents of urban areas may expose to some carcinogenic factors especially those related to the development of multiple myeloma. Previous studies suggested that exposure to engine exhaust, asbestos and benzene may increase the risk of multiple myeloma [8].

Diagnosis of MM is based on the presence of a monoclonal protein, bone manifestations and on bone marrow (BM) plasma cell infiltration. Patients with multiple myeloma must be distinguished from those with monoclonal gammopathy of undetermined significance [< 10% BM plasma cell infiltration, low M-component levels (< 3 g/dl) and no osteolytic bone lesions] and those with amyloidosis or other lymphoproliferative disorders with paraproteinemia. Recent guidelines recommend differentiating between symptomatic and asymptomatic myeloma. Symptomatic patients present with one or more of the CRAB criteria (hypercalcemia, renal failure, anemia, bone lesions) and need active treatment, in contrast to asymptomatic patients, which should be followed only [9].

The disease is relatively rare and the prognosis is poor with a 5-year relative survival of 38.5%. The older age, male gender, black race, family history of the disease and MGUS are all risk factors [10]. The increase in mortality rate has been reported in Japan, Italy, France, Germany, and Wales. Overall, mortality rates are highest among patients older than 85 years. In England and Wales, the mortality rates for men and women aged 70–74 years were higher during the period 1981–1985 compared with 1970–1980, whereas the corresponding rates stabilized over time in the younger age groups [11].

The combination of melphalan and prednisone produces responses in approximately 50% of patients and a disease-free survival (DFS) of approximately 15 months. Meta-analysis comparing combination chemotherapy with melphalan and prednisone has shown no statistically significant difference in survival, despite a higher response rate with more aggressive combination chemotherapy. The VAD regimen (infusional vincristine and doxorubicin combined with dexamethasone) results in a response rate of about 70% and does not compromise stem cell collection unlike melphalan [12].

Thalidomide, an oral immunomodulatory drug, is efficacious for patients with relapsed and refractory multiple myeloma. It has been combined with dexamethasone, and a recent clinical trial noted a higher response rate (70%) with that combination when compared with dexamethasone alone (50%) [13]. More recently, lenalidomide (a thalidomide analog), and bortezomib (a proteasome inhibitor) are evaluated in different combinations with chemotherapy, dexamethasone, or both. Survival information is as yet inconclusive for these combinations [14].

In this retrospective study, we try to review the epidemiological features and survival of PCDs patients diagnosed and treated in the period between 2000 and 2010 and compare our results with other studies.

Patients and methods

Study population

The current study was carried out on 217 Egyptian patients with PCDs. Patients were chosen during the period between "2000 and 2010" among cases referred to the clinical oncology department, Cairo University. The research was approved by the IRB of the clinical oncology department, Cairo University. They were 128 males and 89 females. Their ages ranged between 27 and 80 years with a mean age of 58.5 years and median of 53.5 years.

All patients were subjected to: *1. Routine Laboratory Tests including*, Complete blood count with differential count, complete metabolic panel (calcium, albumin, and creatinine) and coagulation testing. *2. Myeloma-Specific Testing including*, serum protein electrophoresis, monoclonal protein analysis by immunofixation, urine protein electrophoresis, serum β2-microglobulin, CRP, and LDH, BM aspirate and biopsy, flow cytometry (CD38 & CD138) and *3. Skeletal bone survey including*, plain X-ray films of the spine, pelvis, skull, humeri, and femurs.

Prognostic criteria for MM were applied according to the International Staging System (ISS) (Table 1), which provides two advantages over the traditional Durie–Salmon system. The ISS relies on widely available laboratory parameters and allocates patients to equally sized patients groups with