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Myelofibrosis in a 24-Year Old Nigerian

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Abstract
Myelofibrosis (MF) is a chronic myeloproliferative disorder that is uncommon and for which transfusion of red cells is an integral part of management. Treatment with cytoreductive agents, corticosteroids and androgens has so far been unrewarding. Stem cell transplantation may improve treatment outcome. We hereby report MF in a young Nigerian illustrating the diagnostic and therapeutic difficulties encountered in the management of myelofibrosis in resource poor settings.

Key words: Myelofibrosis, uncommon

Résumé
La myélofibrose est un désordre myéloprolifératif chronique rare dont la transfusion des globules rouges fait partie intégrante de sa prise en charge. L'utilisation des agents cytoréductifs, des corticostéroïdes et d'androgènes a jusqu'à là donné que des résultats décevants. La transplantation des cellules à l'origine d'un clone (précurseur) pourrait améliorer le résultat du traitement. Nous rapportons ici le cas d'une myélofibrose chez un jeune nigérian illustrant les difficultés diagnostiques et thérapeutiques rencontrées dans la prise en charge de cette maladie dans un contexte de ressources économiques limitées.

Mots-clés: Myélofibrose, rare

Introduction

Myelofibrosis (MF) is a chronic, progressive often lethal myeloproliferative disease.[1, 2] It is characterised by prominent bone marrow stromal reaction including collagen fibrosis, osteosclerosis and neoangiogenesis, which result from deregulation of myeloproliferation.[3, 4] Males are more affected than females in a ratio of 1.4:1.[5] The median age of presentation is 60.7 years, while persons younger than 30 years account for 2.8% of all cases.[1] Clinical features include, cachexia, constitutional symptoms, transfusion dependent anaemia, splenomegaly, tear drop poikilocytosis, and leucoerythroblastic blood films.[2,6,7] Varying degrees of perivascular marrow fibrosis have been observed in persons with sickle cell disease.[8] Cytogenetic changes have been reported in 38% of Japanese patients.[5] Treatment aims at restoring erythropoiesis using corticosteroids, androgens, hydroxyurea, busulphan, growth factors, cyclosporine, and interferon.[2,3,9] Thalidomide that has been found to reduce anaemia and thrombocytopaenia is an investigational drug. Palliative measures that improve the quality of life are splenectomy, splenic irradiation, and measures that block tumour necrosis factor-α that cause constitutional symptoms.[3] Allogeneic stem cell transplantation has improved survival with a median value of 3.5 -5 years after diagnosis.[1-3, 10]

Case Report

A 24-year old university student was referred with a two-month history of body weakness, fever, haematochezia and weight loss. He had received 6 units of red cell concentrates. He denied exposure to insecticides and petroleum products. Viral serology was not contributory. His haemoglobin electrophoretic pattern was AA. Physical examination revealed a moderately wasted young man with severe pallor and pyrexia of 39ºC. The spleen was 6 cm below the left costal margin. The haematocrit was 18%, leucocyte count 5.3 x 10^9/l, and platelet count 89 x 10^9 /l. His reticulocyte count was <0.0001%. There were some tear drop erythrocytes in his blood film with 4 normoblasts per100 leucocytes interspersed by myelocytes. All bacteriology and chemistry results were not remarkable. The bone marrow aspirate yielded nothing, while the bone marrow biopsy showed replacement of normal haemopoietic elements by early fibrosis, which is composed of spindle cells having fibrillary cytoplasmic processes accompanied by an increase in the collagen deposition interspersed with reticular

fibres on a background of normal iron stores. Ferrokinetic and cytogenetic studies were not done. Therapeutic plan included nandrolone, interferon, and methylprednisolone along side red cell transfusion. He received 40mg of methylprednisolone per day for three weeks without a rise in the reticulocyte count. Three months later he sought discharge from the hospital against medical advice.

Discussion

The presentation of Myelofibrosis in persons less than 40 years old is uncommon. The haemoglobin electrophoretic pattern of AA, has excluded sickle cell disease as a cause of MF. Massive splenomegaly was absent in our patient. The presence of constitutional symptoms is in agreement with the observation of Mesa et al.[2] Therapeutic measures were limited to counselling, red cell transfusion, corticosteroids and anabolic steroids that were in administered in sub optimal doses. This is contrary to the practice of using busulphan, hydroxyurea, interferon-α, stem cell transplantation among others. [2, 3, 9]

Unlike the Japanese patients, [5] and those reported by Hoffbrand, [6] our patient did not undergo cytogenetic and ferrokinetic studies thus illustrating the difficulties encountered in the management of MF in developing countries. The request for discharge against medical advice prompts the need for the introduction of hospice facilities. Hospices reduce the burden of caring for such patients, while maintaining contacts with health care facilities. Our patient belongs to the minority for which there is a need to introduce stem cell transplantation facilities with a view to improving the outcome of managing MF and other stem cell disorders.[1-3, 10]

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