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Short Communication

Heavy chain disease in non-Ashkenazi or Sephardi Jews!

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Abstract

Heavy chain disease is found in Non-Ashkenazi or Sephardi jews.

Studies of people with heavy-chain disease have revealed an association with Sephardi Jews. The gamma heavy chain has been implicated in the disorder. The gamma heavy chain is linked to the Fc fragment of the heavy chain.

Keywords: sephardi jews; immunoglobulin molecule

Introduction

Heavy chain disease is found in Non-Ashkenazi or Sephardi jews.

Studies of people with heavy-chain disease have revealed an association with Sephardi Jews. The gamma heavy chain has been implicated in the disorder. The gamma heavy chain is linked to the Fc fragment of the heavy chain.

Kappa and lambda are the so-called light chains of the immunoglobulin molecule.

These patients may present with Bence Jones protein in their urine.

Heavy chain disease may p [resent with the full gambit of multiple myeloma viz., osteolytic lesions but the IgD varietry may have sclerotic zones.

Typically, the IgA variety has a flame appearance to the abnormal plasma cells and typically the perinuclear halo and cart-wheel appearance of the cell is lost.

Salmon and Drury stage the heavy chain disorder from I to IV depending on the dgree of invasion and the extent of invasion.

A plasma cell/myeloma cell line with specific heavy chain disease characteristics has been established to determine the Heavy chain locus. Among Sephardi or non-Ashkenazi jews the inheritance is autosomal recessive and affects approximately 2.4 % of the population.

Treatment is with pulsed therapy consisting of melphalan and cyclophosphamide until there are no myeloma cell in the bone marrow aspirate or trephine. Classically on treatment the very elevated erythrocyte sedimentation rate comes down.