The Effectiveness of Deferiprone in Thalassemia

To the Editor:

In a note added in proof to their excellent review on iron chelation, Olivieri and Brittenham updated your readers on two abstracts recently presented by them to suggest that deferiprone may not provide adequate control of body iron in a substantial proportion of patients with thalassemia major. Because reviews aim at allowing the reader a balanced view on an issue, it is of importance to inform your readership that the same data have been interpreted differently by other investigators involved in these studies.

More work is needed before one can conclude on the relative efficacy and effectiveness of deferiprone as compared with deferoxamine, as well as on the mechanisms of variability in its response. For example, it has been shown that more sustained administration of deferiprone induces higher iron excretion, and the development of sustained release formulation should be considered.

Future studies should also address potential combinations of deferiprone with deferoxamine in patients exhibiting poor compliance or severe toxicity with desferoxamine. It is worth remembering that it took almost 15 years between the introduction of deferoxamine and the establishment of an effective way to deliver it to chronically iron overloaded patients.

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REFERENCES


Serum Ig Abnormalities in Mantle Cell Lymphoma

To the Editor:

Although the term mantle cell lymphoma (MCL) was coined only in 1982, it is now recognized as a distinct entity with accurate diagnostic criteria [characteristic cytopathic and pathologic features; coexpression of CD5 and B-cell markers, including bright staining for monoclonal surface Ig generally without CD23 expression; and a t(11;14)(q13;q32) that is found in approximately 40% of cases and juxtaposes the bcl-1 gene and the J region of the Ig heavy chain locus]. In their recent thorough review of MCL, Weisenburger and Armitage briefly mentioned hypogammaglobulinemia and monoclonal gammopathy as being decidedly uncommon and serum Ig were not even alluded to in two other recent reviews. Our experience is different, because we have observed a high incidence of such serum Ig abnormalities.

Diagnosis of MCL in the 42 patients (33 men and 9 women; 32 to 88 years of age; mean, 62.5 years of age; 33 untreated and 9 having received chemotherapy) observed in our institution was based on histopathologic features of lymph node biopsies, cytotologic studies of blood, bone marrow and/or lymph node prints (independently reviewed by two of us [G.F. and A.B.]), and immunophenotyping (performed in 35 cases) showing CD5 (34 cases) B cells that strongly stained for monoclonal surface Ig and were negative for CD23 in 20 of 23 cases studied. Cytogenetic analysis showed a t(11;14)(q13;q32) in 20 of the 40 cases studied (50%). Serum monoclonal Ig (moIg) were characterized by thin-layer agarose electrophoresis and immunoelectrophoretic (IEL) analysis and confirmed in some cases by a sensitive immunoblotting procedure. Only those moIg that were detectable by IEL were taken into account, because moIg evidenced by immunoblotting are common in elderly subjects and are mostly unrelated to the proliferating clone in chronic lymphocytic leukemia. Serum Ig class levels were measured by laser nephelometry. Normal levels determined in normal subjects of various age and sex groups are 6.9 to 14.0 mg/mL for IgG, 0.7 to 4.1 mg/mL for IgA, and 0.34 to 2.4 mg/mL for IgM in adults.

Serum moIg detectable by IEL were found in 10 patients (25.6%), including 8 men and 2 women and 44 to 84 years of age (mean, 63.7 years of age), which is not different from the age of patients without moIg. In 1 patient, the moIg was detected only when the disease relapsed after chemotherapy, whereas the first study of the other patients’ serum evidenced the moIg in the other cases (at diagnosis in 7 cases). These moIg were 8 IgM, 5 ξ and 3 λ, and 2 IgG, one of each light-chain type. They were present in moderate amounts, ie, less than 10 mg/mL, in all but 1 case (13 mg/mL). One of these patients’ sera contained a type II mixed cryoglobulin made up of the monoclonal IgM and of polyclonal IgG. Data on surface Ig were available in 8 of the 10 patients with serum moIg. The isotypes of serum and surface moIg matched in the 6 cases with monoclonal serum IgM. In contrast, the patient with a serum IgG had surface IgM and the lymphoma cells from the patient with a