CORRESPONDENCE

SYSTEMIC CAPILLARY LEAK SYNDROME

To the Editor:

We report another case of systemic capillary leak syndrome associated with monoclonal gammopathy and multiple myeloma. The 68-year-old man had his first episode of shock in February 1992, showing the typical features of the syndrome, as described by Clarkson et al (1), Amoura et al (2), and others, including shock with increase in hematocrit, leukocytosis, a marked decrease in total serum protein and albumin concentrations without proteinuria, generalized edema sparing the lungs, and a monoclonal gammopathy (immunoglobulin [Ig] G kappa and light chains Ig lambda). The bone marrow showed a discrete plasma cell infiltration. There were possible lytic bone lesions in the skull and the vertebra. After his second episode of shock 1 month later, bone marrow showed a moderate infiltration by atypical plasma cells that was diagnostic of multiple myeloma. After his third episode of shock 9 weeks later, he was treated with melphalan and prednisolone, although the myeloma was in Salmon and Durie stage I, under the assumption that the development of systemic capillary leak syndrome might be associated with the paraproteinemia. Several days after the first cycle of chemotherapy the patient had another severe episode of shock. Chemotherapy was continued until the 6th cycle in December 1992. The patient remained free of episodes of shock until February 1994, when another severe attack occurred. Multiple myeloma was still in stage I. After another severe attack of shock (myeloma now in stage II with multiple osteolysis), chemotherapy with melphalan and prednisolone was resumed and continued until September 1996, when progression of plasmacytoma with increasing serum levels of the monoclonal IgG paraprotein and a bone marrow infiltration by plasma cells of 80% to 90% was noted. The patient had four very mild attacks of hypotension between October 1995 and September 1996. He died in December 1996, almost 3 years after the diagnosis of systemic capillary leak syndrome and multiple myeloma, from complications of the multiple myeloma.

We believe that our patient's course was benefited by our experience in managing shock with careful volume therapy, by the patient's own awareness of early signs of shock and prompt presentation at the hospital, and by the early institution of a chemotherapy for multiple myeloma. Amoura et al (2) also reported that no attacks of shock occurred in 2 patients with multiple myeloma after the onset of chemotherapy. We believe that—although not supported by experimental studies (3,4)—the reduction in the serum level of paraprotein by chemotherapy should also be considered in patients with recurrent episodes of systemic capillary leak syndrome without overt multiple myeloma.

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The Reply:

As Beermann et al state, we observed 2 patients with systemic capillary leak syndrome who did not have further episodes of shock after the onset of chemotherapy for multiple myeloma (1). Their observation and ours raise the question whether chemotherapy could be a good treatment for this syndrome. Despite these observations, no conclusions can be drawn from such a limited sample. Beerman et al suggest that chemotherapy might act by reducing the serum level of paraproteinemia. We believe that this suggestion has to be taken with caution. First, it is not clear whether chemotherapy or evolution of systemic capillary leak syndrome into multiple myeloma was responsible for improvement. Second, as we discussed previously (2), there is no clear demonstration that the monoclonal immunoglobulins are directly involved in the occurrence of shock. Other mechanisms, such as cytokine secretion by the plasma cell clone, could explain the shock and the possible efficiency of chemotherapy. Third, plasmapheresis that induces a marked reduction in serum paraprotein levels is not effective in our experience (1).

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