To the Editor:

I found the case report by Martinelli et al, “An Unusual Heart Failure: Cardiac Amyloidosis due to Light Chain Myeloma,” interesting, but, unfortunately, I believe that the authors have perpetuated a common misperception in describing the patient as having smoldering myeloma. Careful examination of the data presented indicates that their patient did not have multiple myeloma, and I believe that their conclusion that “light chain myeloma—may be a cause of cardiac amyloidosis” is semantically incorrect.

Light chain (AL) amyloidosis is a plasma cell dyscrasia in the same family as, but distinct from, multiple myeloma. Classical myeloma is characterized by bony lesions, hypercalcemia, and markedly elevated circulating paraprotein, whereas smoldering myeloma is limited to increased monoclonal plasma cell infiltration of the bone marrow without systemic features other than a circulating paraprotein. Although classical myeloma may coexist with, or precede, immunoglobulin-derived amyloid deposition in vital organs, the patient presented had none of the features of myeloma, but did show all the features of pure AL amyloidosis. By virtue of having amyloid deposition in the heart, she cannot, by definition, have had smoldering myeloma. In contrast, AL amyloidosis is associated with an increase in bone marrow plasma cells (occasionally >20%), elevated free λ or (less commonly) κ light chains, low-level (or, as in this case, sometimes undetectable) serum monoclonal immunoglobulin band, and documented organ deposition of amyloid. On the basis of the data presented, this patient had AL amyloidosis alone, without myeloma. Furthermore, in cases where myeloma and amyloidosis coexist, it is not the light-chain myeloma that causes amyloidosis; rather, both diseases are different manifestations of a single underlying process, namely, a plasma cell dyscrasia.

AL amyloidosis in any organ, particularly the heart, can progress rapidly and has a high morbidity and mortality. It almost always justifies aggressive treatment, even when detected very early. In contrast, smoldering myeloma is commonly an indolent disease, and treatment is generally not indicated at the time of diagnosis. Failure to recognize that a low-grade plasmacytosis in the bone marrow can represent the source of the precursor protein for AL amyloidosis can mislead the clinician into ignoring associated clinical features that may represent amyloid deposition, thereby delaying not only the correct diagnosis, but also timely treatment. Thus, the distinction between coexisting myeloma with amyloidosis and pure AL amyloidosis with a typical bone marrow appearance of plasmacytosis is not only semantic, but has extremely important therapeutic considerations.

Disclosures

None.

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References

Letter by Falk Regarding Article, "An Unusual Heart Failure: Cardiac Amyloidosis Due to Light Chain Myeloma"
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