THE AMERICAN JOURNAL of MEDICINE®

A Case of Disseminated Systemic Light-Chain Amyloidosis with Coagulopathy

To the Editor:

In a recent diagnostic dilemma, Ornoff et al[1] describe an interesting case of disseminated systemic light-chain amyloidosis, presenting with predominant liver and renal involvement and a coagulopathy refractory to plasma transfusion. Assuming acquired factor X deficiency was the main cause of this patient's coagulopathy, it is not surprising that fresh frozen plasma contained an insufficient concentration of factor X to correct the coagulopathy. Instead, the use of prothrombin complex concentrate, which contains higher concentrations of factor X, or high-purity Factor X concentrate, would be more likely to temporarily correct the coagulopathy during invasive procedures or to treat major bleeding.[2] It is also important to note that, although this patient declined active therapy of his amyloidosis due to the disease's "overall poor prognosis", there has, in fact, been marked improvement in the prognosis of disseminated systemic light-chain amyloidosis over the past 40 years, in part due to the introduction of effective therapies such as proteosome inhibitors, immunomodulating drugs, and anti-CD38 monoclonal antibodies.[3] In addition, translocation t(11;14) is seen in up to 62% of patients with disseminated systemic light-chain amyloidosis,[4] and the oral BCL-2 inhibitor venetoclax appears to be highly efficacious in such cases,[5] and can be used successfully even in dialysis-dependent patients.[6]

Philip Murphy
Department of Haematology,
Beaumont Hospital, Dublin, Ireland

https://doi.org/10.1016/j.amjmed.2022.06.013

References

1. Ornoff DM, Balaban E, Major A, Berry A, Young E. The missing factor (s): disseminated amyloidosis causing coagulopathy refractory to plasma transfusion. *Am J Med* 2022;135(6):721–3.
2. Dejhansathit S, Suvannasankha A. Acquired factor X deficiency in patients with primary light chain amyloidosis. *J Investig Med High Impact Case Rep* 2019;7:2324709619832332.
3. Staron A, Zheng L, Doros G, et al. Marked progress in AL amyloidosis survival: a 40-year longitudinal natural history study. *Blood Cancer J* 2021;11(8):139.
4. Bochtler T, Hegenbart U, Kunz C, et al. Translocation t(11;14) is associated with adverse outcome in patients with newly diagnosed AL amyloidosis when treated with bortezomib-based regimens. *J Clin Oncol* 2015;33(12):1371–8.
5. Hasib Sidiqi M, Al Saleh AS, Leung N, et al. Venetoclax for the treatment of translocation (11;14) AL amyloidosis. *Blood Cancer J* 2020;10(5):55.
6. Swan D, Delaney C, Natoni A, O'Dwyer M, Krawczyk J. Successful venetoclax salvage in the setting of refractory, dialysis-dependent multiple myeloma with t(11;14). *Haematologica* 2020;105(3):e141–3.

Funding: The author has no relevant funding sources to report.
Conflicts of Interest: None.
Authorship: Philip Murphy was the sole contributor to the manuscript.

Requests for reprints should be addressed to Philip Murphy, Department of Haematology, Beaumont Hospital, Beaumont Rd., Dublin 9, Ireland.
E-mail address: philipmurphy@beaumont.ie