

新たな疾患概念としての IGLC2由来ALアミロイドーシス ～形質細胞性疾患での従来型AL, AH, まれなAAアミロイドーシスとの差異とは～

KEY WORDS

- ALアミロイドーシス
- M蛋白
- 軽鎖

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Abstract

Amyloidosis is the disease that insoluble protein deposits to various tissues. In AL amyloidosis, immunoglobulin light chain (LC) deposits as amyloid protein. Immunostaining is regularly utilized to confirm the diagnosis of AL amyloidosis by proving LC in amyloid lesions. However, there are substantial cases having difficulty in confirming diagnosis despite presence of monoclonal LC in serum. We analyzed those cases by using combination of liquid chromatography and mass spectrometry (LC-MS/MS) to detect causal protein in amyloid lesions. We found that immunoglobulin constant region 2 (IGLC2) was frequently detected. Moreover, immunostaining using anti-IGLL5 antibody, predictably cross-reacts to IGLC2, showed positive staining. These results suggest that IGLC2 often deposits as amyloid protein when LC was not detected by routine immunostaining. We propose that utilization of anti-IGLC2 antibody should contribute for improvement of diagnostic sensitivity of AL amyloidosis, especially for cases with deposition of lambda type LC.

AL amyloidosis caused by
IGLC2 : A new clinical entity.
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