Immunoglobulin Light Chain (AL) Amyloidosis Preceding Marginal Zone Lymphoma: A Case Report

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BACKGROUND

- Amyloidosis is subtyped by being either primary (AL), secondary (AA), or familial (transthyretin amyloidosis (ATTR))
- Mucosal-associated marginal zone lymphoma (MALT)-associated amyloidosis, MALT preceded the diagnosis of amyloid by a median of 64 months

CASE PRESENTATION

- A 54-year-old female with medical history of AL amyloidosis with kappa-restricted plasma cells initially diagnosed in 2011, 11 years prior to the current presentation
- During the time of her diagnosis, 11 years prior, she had no evidence of cardiac involvement based on a cardiac MRI and her renal function was normal.
- Lost in follow-up and did not receive medical care until November 2020, when she presented to an outside hospital with acute renal failure. Subsequently, she was diagnosed with systemic amyloidosis with renal involvement
- Transferred to outside hospital for management of 17cm left groin mass. Biopsy results showed infiltrate of small lymphocytes positive for CD20 and CD79a, consistent with MZL
- She was started on daratumumab, bortezomib, and dexamethasone, which she continues to the present time on an ambulatory basis

DISCUSSION AND CONCLUSION

- This case is unusual as the diagnosis of systemic AL amyloidosis preceded that of lymphoma by several years
- Average survivorship is 26 months with this diagnosis. She responded well to the therapies, which is unusual, and has had an extended survival response since receiving treatment
- One aspect that deserves investigation is whether the AL amyloidosis and MZL arose from a single pre-B cell or B cell clone or represent clonal differentiation of a B cell progenitor among two allied by divergent pathways
- The MZL expressed both kappa and lambda light chains suggestive of differentiation along divergent pathways.

REFERENCES

Figure 1: AL amyloidosis and its associated complications

Figure 2: A: Showing the aggregate of lymphoid cells, which are small in size with moderate pale cytoplasm associated with some plasma cells. B: CD20 cells. C: CD5 cells. D: Multiple lymphoid aggregates associated with extensive areas of pink amorphus martial deposition. E: Congo red showing positive brick red staining of the abnormal material. Congo red under polarized light showing apple-green birefringence. F: Abnormal material stain positive with crystal violet.

UNIQUENESS

- It rarely occurs in patients with low-grade lymphoproliferative disorders and even more rarely in patients with marginal zone lymphoma (MZL) without mucosal involvement
- Patient diagnosed with systemic amyloidosis with renal involvement 11 years prior to the diagnosis of MZL