Adult Hematology Abstract Categories

Myeloma
OP 06

A TALE OF THE CRUMBLING AMYLOID WALL: BREAKING THROUGH LIVER STIFFNESS IN AL AMYLOIDOSIS

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Objective: Systemic AL amyloidosis is characterized by the deposition of misfolded amyloid fibrils in tissues, produced by clonal plasma cells. Daratumumab, a human IgG kappa-type monoclonal anti-CD38 antibody, is expressed on the surface of plasma cells, hematopoietic stem cells, regulatory T and B cells, monocytes, and dendritic cells. Due to this broad range of activity, daratumumab is thought to improve organ responses in AL amyloidosis through direct effects on tumor cells and via immunomodulatory mechanisms, providing additional therapeutic benefits. FibroScan, a noninvasive method for measuring liver stiffness, is routinely used today in the diagnosis and follow-up of chronic liver diseases. One condition that increases tissue stiffness is the accumulation of amyloid deposits in tissues. Various studies suggest that it can be useful in diagnosis when tested for this purpose. We aim to present two cases demonstrating that FibroScan can be a valuable tool not only in diagnosis but also in monitoring the progression of the disease. **Case report:** Case 1 A 53-year-old male patient was referred to our clinic after an incidental finding of M protein (2 g/dl) on serum protein electrophoresis requested at the Neurology Clinic, where he was being followed for epilepsy. An IgG lambda-type paraproteinemia was documented by immunofixation electrophoresis. His tongue was slightly noticeably enlarged. With suspicion of AL amyloidosis abdominal fat aspiration was performed, which revealed amyloid existence by Congo red stain positivity. He has no other organ involvement symptoms and signs but the liver was greater than normal size being 17 cm in the mid-clavicular line. The serum alkaline phosphatase (ALP) level was within normal limits. The liver elastography (FibroScan) result was 9 kPa, consistent with moderate scarring. He was monitored without intervention. Three years later when he developed peripheral neuropathy-related symptoms, AL amyloidosis management was decided. The liver size was similar and serum ALP level was still normal but the FibroScan result changed to severe scarring as being 9.1 kPa. Case 2 A 60-year-old female patient presented with a 20 kg weight loss over one year and chest pain. Imaging revealed diffuse infiltrates in the lungs. A bronchoscopy biopsy was consistent with AL amyloidosis. The patient had parenchymal lung involvement, anemia (Hb 9.7 g/dL), and an interventricular septal diameter (IVSD) of 1.5 cm. She was classified as Mayo 2012 stage 1 and Palladini renal stage 1. At diagnosis, her ALP level was 308 IU/L (laboratory upper limit: 130 IU/L) and her Fibroscan result was 51.2 kPa. The patient was started on daratumumab, bortezomib, cyclophosphamide, and dexamethasone therapy. After 18 months of monthly daratumumab treatment, follow-up measurements showed an ALP of 221 IU/L and a Fibroscan of 9.1 kPa. During the same period, hematological response was assessed as a very good partial response (dFLC: 19.6 mg/L), and NT-pro BNP decreased from 1558 to 512 pg/ml **Conclusion:** In the first case, liver stiffness measurements remained nearly stable over two years with a slight increase during clinical progression. In the second case, despite stage I cardiac and renal involvement, liver stiffness was very high and showed a striking reduction after treatment. Thus, liver stiffness may be an occult sign of liver involvement and may provide insights for monitoring disease progression

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OP 07

A SINGLE-CENTER REAL-LIFE EXPERIENCE WITH FIRST-LINE DARATUMUMAB, BORTEZOMIB, CYCLOPHOSPHAMIDE, AND DEXAMETHASONE (DARA-VCD) IN AL AMYLOIDOSIS

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Case Report: Systemic amyloidosis results from the production of misfolded immunoglobulin light chains by monoclonal CD38 + plasma cells. These misfolded light chains form amyloid fibrils, which accumulate in various tissues and cause organ damage. Following the results of the Phase 3 ANDROMEDA study, where the addition of daratumumab, an anti-CD38 agent, to first-line treatment showed favorable outcomes, Dara-VCD has become a standard first-line therapy. In this study, we compared the outcomes of patients with AL amyloidosis who were treated with first-line Dara-VCD in our clinic to those treated with other triplet regimens. **Methodology:** Patient's data with AL amyloidosis followed between 2010 and 2024 were retrospectively reviewed from the institution's database. Two groups were established patients treated with Dara-VCD and those without Dara. The clinical characteristics and response criteria were compared using SPSS 21. **Results:** A total of 52 patients were included in the study, with a mean age of 60 ± 10 years for the entire group. There was no statistically significant difference in the demographic distribution between the groups ($p = 0.003$). The median follow-up period was 32 months (1-114 months). In 27 (51.9%) patients, cardiac involvement was present, and 26