St Lukes

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Introduction

- Autoimmune hepatitis (AIH) is a chronic inflammatory liver disease that typically requires lifelong immunosuppression to prevent fibrosis.
- We present a patient with type 1 AIH who achieved biochemical remission following autologous stem cell (SC) transplantation for IgA multiple myeloma.

Case

- A 67-year-old female presented with abdominal pain and jaundice.
- Labs revealed elevated liver enzymes with an elevated ANA and anti-smooth muscle antibody.
- Type 1 AIH was confirmed by liver biopsy (A & B), which revealed stage 2 fibrosis according to the Ludwig-Batts classification.
- She was started on prednisone and azathioprine. Steroids were able to be withdrawn, but she remained on azathioprine and Ursodiol for years due to recurrent exacerbations.
- 2 years later, she was diagnosed with multiple myeloma confirmed on bone marrow biopsy (C & D).
- Prednisone, Ursodiol, and azathioprine were all discontinued, and the patient underwent an autologous SC transplantation.
- She has since been continued on maintenance ixazomib and low dose dexamethasone once weekly but has been monitored off her previous AIH therapies.
- 2 years following SC transplantation, she remains in biochemical remission with normalization of her liver enzymes.

Images



(A) H&E stain of the liver showing portal tract with artery (red arrow), bile duct (yellow arrow) and interface hepatitis (blue arrow) comprised of numerous plasma cells and scattered lymphocytes. (B) H&E stain of the liver showing lobular inflammation consisting of lymphocytes. (C) H&E stain of the bone marrow showing cellular marrow with numerous plasma cells (blue arrows). (D) Bone marrow, Kappa in situ hybridization reveals Kappa-light chain restricted plasma cell neoplasm.

Discussion

- Treatment options for AIH remain limited to immunosuppressant medications like corticosteroids and azathioprine, which can lead to substantial adverse side effects.
- Although limited data exists and additional clinical investigations are required, many have postulated the effects of SC transplantation in patients with AIH given their promising results in animal studies.
- Therefore, our case encourages further studies investigating the use of stem cells as an alternative treatment for AIH or as a synergistic therapy in patients whose AIH remains uncontrolled.

References

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