Assessment of Renal Outcome Following Therapy in Monoclonal Immunoglobulin Deposition Disease (MIDD): A Retrospective Study Highlighting the Need for Consensus Criteria

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Introduction

- Randall-type Monoclonal Immunoglobulin Deposition Disease (MIDD) is a rare condition in which organ dysfunction arises from monoclonal heavy and/or light chains deposition along basement membranes, and predominantly affects the kidneys. MIDD is among the most common subtypes of monomonal gammopathies of renal significance (MGRS) and is associated with plasma cell and lymphoid neoplasms.
- Hematologic responses are characterized according to IWGMM International Myeloma Working Group (IWGMM) consensus criteria.
- Although two renal response systems exist for plasma cell disorders, the International Myeloma Working Group (IMWG) consensus criteria. Hematologic responses are characterized according to IMWG consensus criteria.

Methods

- Objectives

Introduction

- Results

- Methods

Results

- Results (continued)

Conclusions

- References

Fig. 1: A. Renal Survival; B. Renal Response

Table 1: Derived Response Rate per IWGMM/NM Uniform Response Criteria

Table 2: Renal Response Criteria

Table 3: Comparison of Responses by IWG vs. myocardial Criteria

Materials and Methods

- Assessment of Renal Outcome Following Therapy in MIDD using modern plasma-cell directed therapy and evaluate hematologic and renal response to therapy according to existing response criteria.

Results

- Results (continued)

Conclusions

- With most pts treated with bortezomib and autologous stem cell transplantation (ASCT), 26 of 28 (94%) achieved ≥VGPR
- We demonstrate that both IWGMM (based on eGFR) and amyloidosis (based on proteinuria) criteria are needed to capture RR: Among 28 pts whose RR could be assessed, initial renal presentations included proteinuria with preserved eGFR (n=21%, 2), proteinuria and decreased eGFR (n=32%, 9), and decreased eGFR without proteinuria (n=13, 46%).
- Using both criteria, which were concordant in pts with both decreased eGFR and proteinuria, 22 of 28 pts (79%) had a RR, including 2 of 7 discontinuing dialysis. All 6 pts (100%) with isolated proteinuria and 7 of 13 (54%) with isolated decreased eGFR achieved RR, suggesting that isolated proteinuria may be an early and reversible manifestation of MIDD.
- Baseline eGFR was predictive of RR (p=0.02 by quartile), while hematologic response (CR vs. non-CR) was not, probably due to high hematologic response rates, hindering the ability to detect such association.
- With a median-follow up of 110 months (95% CI: 71-119), the overall survival was 136 months (95% CI: 79-119) and median RS has not been reached.

Results

- Results (continued)

Conclusions

- We have used a systematic approach to assess RR in MIDD, a field that remains mired in uncertainty in the literature.
- We show that IWGMM and amyloidosis response criteria are both essential to adequately assess the RR in MIDD. We also show that the RR rate is high and durable in this disease with bortezomib-based treatment and ASCT.
- This study will help inform the development of consensus renal response criteria that are needed in MIDD.

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