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Objective: Isatuximab (Isa, anti-CD38 monoclonal antibody) is approved in combination with carfilzomib (K) and dexamethasone (d), for relapsed multiple myeloma (MM) patients (pts) after ≥1 prior therapy. Final progression free survival (PFS) analysis after 2 years showed mPFS of 35.65 mo (Isa-Kd) vs 19.15 mo (Kd). Here, we report the final overall survival (OS) from IKEMA planned 3 years after the primary PFS analysis. **Methodology:** Pts with 1–3 prior lines of therapy were randomized 3:2 to receive Isa-Kd (n=179) or Kd (n=123). Treatment (tx) was given until progressive disease, unacceptable toxicity, or pt wish. Safety was assessed in all treated pts. **Results:** As of 7 Feb 2023, 23.5% (Isa-Kd) and 5.7% (Kd) pts were on tx. Median follow-up: 56.61 mo. OS benefit was more in Isa-Kd pts (mOS was NR; [95% CI: 52.172–NR] vs 50.6 mo [95% CI: 38.932–NR]; HR: 0.855; nominal one-sided p=0.1836). Isa-Kd had longer TTNT vs Kd (median 43.99 vs 25.0 mo; nominal one-sided p=0.0002), as was PFS2 (median 47.18 vs 32.36 mo; nominal one-sided p=0.0035). The safety profiles were comparable to interim and final PFS analyses. Grade ≥3 TEAEs: 84.2% (Isa-Kd) vs 73.0% (Kd). **Conclusion:** This final OS analysis shows a meaningful trend for OS benefit with Isa-Kd vs Kd despite subsequent tx with anti-CD38 agents, introduction of tx with novel mechanism of action among further therapies, and the COVID-19 pandemic. Improvements in TTNT and PSF2 were observed and sustained PFS benefit still observed at PFS2. The Isa-Kd safety profile was consistent with previous analyses, supporting it as a standard-of-care therapy for relapsed MM pts.

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OP 05

REAL-WORLD (RW) TREATMENT PATTERNS AND OUTCOMES IN PATIENTS (PTS) WITH RELAPSED/REFRACTORY MULTIPLE MYELOMA (RRMM) WITH AT LEAST ONE PRIOR THERAPY IN TURKEY

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Objective: Data on RW treatment patterns and outcomes in RRMM Pts who received at least one prior line of therapy (LoT) are lacking outside the US and Europe. This study evaluated RW clinical characteristics, treatment patterns, and outcomes among Turkish Pts who received at least one prior MM-specific therapy. **Methodology:** This retrospective chart review included RRMM Pts who had received at least one prior LoT and initiated a second-line (2L) or third-line (3L) MM-specific treatment regimen between 01-Jan-2015 and 31-Dec-2020. Patients' demographics and clinical characteristics, treatment patterns, and overall survival (OS) were evaluated. **Results:** Of the 107 RRMM Pts initiating 2L treatment, 91.6% experienced symptomatic disease [prominent symptoms: anemia (71.0%); bone lesions (53.3%)]. Table 1 presents other clinical and demographic characteristics. Bortezomib (BOR)-based regimens were most used in first-line (1L) regardless of stem-cell transplant (SCT) status (SCT induction: 68.7%; non-SCT: 79.5%), and lenalidomide (LEN-based regimens were used as 1L maintenance (40.3%). LEN-free regimens were used in 58.1% (2L) and 35.6% (3L) of Pts, with DVd (29.5%) and DRd (19.5%) being the most utilized regimens in 2L and 3L, respectively (Fig. 1). In total, 53.1% were LEN-retreated and 30.8% were LEN-refractory. The median (interquartile range) duration of treatment on 2L [7.0 (6.0, 10.5) months] and 3L [7.1 (6.0, 14.0) months] was short (Table 2). After 2L and 3L initiation, 57.9% and 25.6% of Pts had disease progression; median OS was 10.4 and 12.8 months, respectively (Table 3). **Conclusion:** BOR-based regimens were commonly utilized in 1L. LEN-based regimens were used as maintenance therapy in 1L and as retreatment in RRMM Pts. Newer therapies (Daratumumab- or Carfilzomib-based regimens) were utilized in 2L and 3L. The short duration of therapy, high disease progression rate, high LEN retreatment, and refractoriness rates indicate the need for new LEN-free regimens for treating RRMM Pts in Turkey.

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OP 06

URETERAL AMYLOİDOSİS: A CASE REPORT OF SUCCESSFUL MANAGEMENT WİTH SURGERY, RADİATİON, AND CHEMOTHERAPY

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Background: Ureteral amyloidosis is a unique and infrequent form of amyloidosis characterized by the deposition of amyloid proteins within the ureters. These tubes, responsible for transporting urine from the kidneys to the bladder, can become obstructed due to this protein accumulation, potentially leading to renal complications. We are presenting a case ureteral amyloidosis. **Case Report:** A 48-year-old male with no known prior medical conditions presented with a three-month history of right-sided pain, frequent and painful urination, reduced urine output, and hematuria. Blood tests showed a hemoglobin level of 12.8 g/dL and MCV of 73. Urinalysis revealed pyuria and hematuria. An upright abdominal X-ray indicated hydronephrosis, and an abdominal CT scan

detected grade 2 hydronephrosis on the right side and a suspicious mass in a 1 cm segment of the distal right ureter outside the bladder. The mass was excised, and histological examination with crystal violet and Congo red staining showed a strong positive reaction for amyloid. Immunohistochemical analysis confirmed the diagnosis of lambda light chain amyloidoma. Systemic screening for amyloid deposition was negative except for the ureter. Nine months post-operation, the patient returned with recurrent pain and oliguria. A CT scan revealed a mass at the excision site, consistent with lambda light chain amyloidoma. Considering it a recurrent disease, the patient underwent intensity-modulated radiation therapy (IMRT) with a total dose of 20 Gy in 10 fractions of 2 Gy each. Two months post-radiation, with recurring symptoms, the patient received four cycles of bortezomib-dexamethasone treatment. Post-treatment, the patient's symptoms improved, and CT imaging showed the disappearance of the mass lesion. **Conclusion:** Ureteral amyloidosis, though rare, can present with significant clinical symptoms. Early detection and a combination of surgical and medical interventions, as demonstrated in this case, can lead to symptom resolution and improved patient outcomes.

Keywords:
Amyloidosis
Ureteral Amyloidosis
Bortezomib
Surgery
Radiotherapy

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Adult Hematology Abstract Categories

Platelet Diseases
OP 07

THE ROLE OF ADAMTS13 ACTIVITY LEVELS ON DISEASE EXACERBATION OR RELAPSE IN PATIENTS WITH IMMUNE-MEDIATED THROMBOTIC THROMBOCYTOPENIC PURPURA: POST HOC ANALYSIS OF THE PHASE 3 HERCULES AND POST-HERCULES STUDIES

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Objective: The management of exacerbations and disease relapse is important for patients with immune-mediated thrombotic thrombocytopenic purpura (iTTP). Severe ADAMTS13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13) deficiency during clinical remission is associated with risk of relapse and may guide prophylactic immune-modulatory therapy. We evaluated ADAMTS13 activity as a potential biomarker of exacerbation or relapse risk in the HERCULES and post-HERCULES studies. **Methodology:** This is a post hoc analysis of integrated data from the modified intent-to-treat (mITT) population of the Phase 3 HERCULES trial (NCT02553317) comparing caplacizumab and placebo (both plus standard-of-care treatment) in patients (pts) with iTTP and the 3-year follow-up post-HERCULES study (NCT02878603). ADAMTS13 activity was determined at baseline, weekly during treatment (post-TPE) and twice during follow-up. Recurrence risk was assessed according to ADAMTS13 activity, using TTP adverse event codes. **Results:** 49/144 (34%) pts in the HERCULES mITT had a recurrence during HERCULES or post-HERCULES. 140/144 pts had follow-up data after treatment end. Of these, 39 pts (28%) had a recurrence after treatment end; mean [SD] ADAMTS13 activity was 20.5% (28.7) in pts with recurrence vs 54.0% (34.9) in pts without; [P<0.0001]). ADAMTS13 activity was <20% at treatment end in 69.2% (27/39) and 27.1% (26/96) pts with/without recurrence (P<0.0001). Similar trends were seen across both treatment groups (Table). **Conclusion:** Regardless of the treatment received (caplacizumab or placebo), lower ADAMTS13 activity levels at end of treatment were associated with a higher risk of recurrence in the HERCULES and post-HERCULES studies. These data highlight the predictive value of ADAMTS13 levels on the risk of recurrence and may assist clinical decision-making in the treatment of iTTP. This content was first presented at ASH 2022 (abstract #2493).

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