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
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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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