Use of Avacopan as a Steroid-Sparing Medication in ANCA-associated Vasculitis Patients with Renal Impairment

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Background: Alveolar hemorrhage can be a significant cause of mortality in patients hospitalized with ANCA-vasculitis and will often necessitate use of high-dose glucocorticoids early on in the hospital course. When considering a transition to steroid-sparing medications, options may be limited in patients with renal impairment. This case highlights the use of the C5a receptor inhibitor avacopan as an option in such patient populations.

Case Presentation: A 69-year-old female with a history of CKD stage IIIA, insulin-dependent type 2 diabetes mellitus, congestive heart failure, and hypertension was admitted to the hospital following a 1–2-week history of fatigue and weakness after outpatient lab studies were significant for a hemoglobin of 7.1. The initial chest x-ray showed only mild bibasilar opacities, but over the next couple of days, the patient began to experience worsening respiratory distress, ultimately requiring intubation. Due to significant fluid overload and worsening creatinine, the patient was started on hemodialysis. Bedside bronchoscopy showed erythematous mucosa of the bilateral bronchial trees with multiple foci or oozing blood concerning for alveolar hemorrhage. Rheumatologic work-up was significant for ANA at 1:1280 with homogenous pattern, normal C3/C4, elevated MPO at 260 and PR3 at 36 with a positive p-ANCA. The patient was immediately started on high-dose IV glucocorticoids. When the time came to wean down steroids, cyclophosphamide was considered as an adjunct steroid-sparing therapy. However due to concerns for renal toxicity, a plan was made to start avacopan instead. Prior to initiation of avacopan, the patient's family ultimately decided to pursue comfort measures.

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Conclusion: Since alternative pathway complement activation has been shown to play a role in the pathophysiology of ANCA-associated vasculitis1, pharmaceuticals such as avacopan that inhibit this pathway may be considered as part of the treatment plan. This is especially true in patients with renal impairment, which is a common sequela of ANCA-associated vasculitis.

Keywords: ANCA-Associated Vasculitis, Vasculities, Avacopan

References

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