Hypocomplementemic-urticarial vasculitis syndrome (hvus): A diagnostic dilemma

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Introduction: HVUS Is an extremely rare debilitating condition with reported incidence of 0.5/100000. Presence of recurrent and chronic urticarial rash is the dominant clinical finding in HVUS. Exact pathophysiology is unknown, however, it has been demonstrated that immune-complex mediated injury is the predominant mechanism leading to systemic manifestations just like in SLE. It can involve multiple systems and pulmonary involvement is the leading cause of mortality and morbidity in HVUS. This article serves to create more awareness about this rare condition among clinicians and specifically highlights the appropriate diagnostic strategies and treatment options available, with special emphasis on the latest advancements in management as well as ongoing or recent clinical trials regarding treatment of this condition.

Case Presentation: Our patient was a 47-year-old Caucasian female whose initial symptoms started with episodic urticaria which gradually progressed to respiratory symptoms such as hoarseness, chest wall and laryngeal edema. Patient's symptoms occurred gradually and progressed stubbornly over the past 2 years. Her personal and family history was also unremarkable for any autoimmune or rheumatological conditions. She became increasingly resistant to conventional treatment with antihistamines and steroids. During frequent ED visits, her symptoms were partially responsive to fresh frozen plasma, epinephrine and high-dose steroid therapies given together. Outpatient, medications targeting complement and bradykinin-mediated pathways failed to produce any significant improvement. Blood tests were only significant for low complement levels and positive p-ANCA. She suffered immense stress along with delayed diagnosis and multiple failed treatments and multiple negative skin biopsies, before finally a 3rd skin biopsy, clinched the above diagnosis.

Conclusion: This case serves to highlight the insidious and non-specific nature of this morbid condition along with the common mimics for this condition. The role of skin biopsy is extremely important in the diagnosis along with the utility of multiple skin biopsies. Even after initial negative biopsies, the finding of low complement along with recurrent urticarial rash, should have alerted the clinicians much earlier about this rare condition and would had allowed earlier treatment. Lastly, this case also highlights the

experimental nature of most drugs which are currently employed in treating HVUS. Treatment of HVUS currently usually employs different types of immunosuppressants and nowadays, biologics are being increasingly used to block functionality of different cytokines.

