

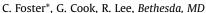
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symptoms. While the current focus in urticaria treatment is with antihistamines and omalizumab, this case demonstrates the role for cyclosporine in treatment of DPU, an option which is not currently described in the literature. ¹

M096

TREATING ANOTHER CASE OF CHOLINERGIC URTICARIA? — NO SWEAT!





Introduction: Cholinergic urticaria is commonly encountered in allergy practice. Its signature pattern of pruritic, pinpoint wheals and surrounding erythema, triggered by heat and exercise, make it one of the most recognizable of the chronic inducible urticarias. However, things aren't always what they seem. Asking one question, in particular, could suggest a more ominous condition with significant treatment implications — "Do you sweat?"

Case Description: A 28-year-old Caucasian male presented to the allergy/immunology clinic for a second opinion on his recalcitrant cholinergic urticaria. He experienced near-daily symptoms despite trials of high dose antihistamines, montelukast, and omalizumab over the previous 2-year period. Further history revealed he had experienced a profound elevation of core body temperature while flying in an aircraft cockpit prior to the onset of his symptoms. Since then, he is no longer able to sweat, even when he exercises or exerts himself, and has since required modification of his daily activities to prevent a flare of his urticaria. Collaborative evaluation with dermatology and confirmatory sweat gland biopsy solidified the diagnosis of acquired idiopathic generalized anhidrosis of the idiopathic pure sudomotor failure subtype. He was treated with high dose pulsed steroids for 5 days with good initial response.

Discussion: Acquired idiopathic generalized anhidrosis (AlGA), reported as case series out of Japan, is rarely diagnosed in the United States. Identifying patients with cholinergic urticaria who concomitantly suffer from anhidrosis may uncover a higher prevalence of AlGA and subsequently direct therapeutic decisions towards improved outcomes for this unique set of patients.

M097

ANGIOEDEMA WITH ACQUIRED C1 ESTERASE INHIBITOR DEFICIENCY ASSOCIATED WITH PAPILLARY UROTHELIAL CARCINOMA



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Introduction: Angioedema with acquired C1 esterase inhibitor deficiency is a rare form of acquired angioedema which generally presents in older patients who have an underlying malignancy, lymphoproliferative, or autoimmune disorder. Other malignancies have been noted in some cases, but there has only been one reported case associated with bladder carcinoma; we present the second.

Case Description: A 57-year-old male presented with facial and lip swelling and severe abdominal cramping. He had no history of laryngeal angioedema, urticaria, use of angiotensin converting enzyme inhibitor (ACE-I), or a family history of hereditary angioedema. Initial C4 level was low, followed by a normal, and then elevated level two and five months later respectively. All subsequent C4 levels remained low. Supporting labs included low C1INH level, C1INH function, and low C1q level, consistent with the diagnosis of acquired angioedema from C1 esterase deficiency. Workup did not reveal a malignancy or autoimmune disease. Eight years after his initial diagnosis, the patient was found to have low-grade papillary urothelial carcinoma after evaluation of acute gross hematuria.

He had recurrence with multiple high-grade noninvasive papillary urothelial carcinoma the following year. Ten years later, he continues to be monitored without recurrence or evidence of other malignancy.

Discussion: The patient's increase in C4 level in the beginning of his evaluation reiterates the importance of following mild abnormalities in C4 level with consistent clinical presentation over time. In addition, the presentation of bladder carcinoma 8 years after initial diagnosis highlights the importance of frequent monitoring with thorough review of systems and physical exam.

M098

A CASE OF HEREDITARY A-TRYPTASEMIA EXACERBATION FOLLOWING SECOND DOSE OF MRNA COVID-19 VACCINE ON OMALIZUMAB



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Introduction: Patients with hereditary α -tryptasemia (H α T) can have multi-organ symptoms that may respond to omalizumab. Whether H α T predisposes to systemic allergic reactions to the mRNA COVID-19 vaccinations is unknown.

Case Description: A 47-year-old woman with hypothyroidism, gastroparesis, drug allergies, and asthma presented with recurrent episodes of anaphylaxis treated with epinephrine associated with elevated tryptase levels (baseline 20.7). Genetic testing showed α tryptase copy number 2 and β -tryptase copy number 3. Bone marrow biopsy was negative for mastocytosis. Treatment with antihistamines, montelukast, steroids, cyproheptadine, and cyclosporine led to relative control of symptoms and omalizumab was started to taper immunosuppressants. She tolerated her first Pfizer COVID-19 vaccine, but developed anaphylaxis after her second dose requiring multiple doses of epinephrine and had recurrence of daily symptoms following this. Four days later, she presented for her third omalizumab injection and developed immediate urticaria, generalized flushing, and throat tightness treated with epinephrine. Given prior tolerance, omalizumab was continued. She had more mild symptoms with subsequent two omalizumab doses that responded to diphenhydramine and famotidine. She has since been pre-medicated with antihistamines, famotidine, lorazepam, and fluids prior to omalizumab with good response and significant improvement in symptoms allowing her to taper off cyclosporine

Discussion: H α T leads to increased risk of idiopathic anaphylaxis which can be challenging to manage. This case showed exacerbation following second dose of mRNA COVID-19 vaccine (despite tolerating first dose) that likely precipitated reactions following omalizumab. Therefore, omalizumab was able to be safely continued with pre-medication and was effective in controlling symptoms.

M099

WORSENING ANGIOEDEMA IN A PATIENT WITH A DIAGNOSIS OF HEREDITARY ANGIOEDEMA (HAE)



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Introduction: Acquired angioedema (AAE) is a rare disorder and can be challenging to diagnose with complement level alone. This case highlights the importance of history in guiding the diagnosis of AAE.

Case Description: The patient is a 40-year-old male without a clear family history of angioedema who has been diagnosed with HAE three years ago at an outside clinic based on low C4 (<4 mg/dL), C1INH protein (17 mg/dL) and function level (11%). C1q was normal