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A CASE OF A WOMAN WITH RELAPSING POLYCHONDRITIS, CELIAC DISEASE, AND IMMUNOGLOBULIN A DEFICIENCY

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Introduction: Relapsing polychondritis (RP) is a rare immune-mediated disease characterized by episodic inflammation of cartilage and other tissues in the body. Many patients with RP have concurrent diseases, commonly other rheumatologic conditions. We describe a case of a woman with RP, Rheumatoid arthritis, Celiac disease, selective IgA deficiency, recurrent infections and asthma.

Methods: Case Description.

Results: A 21-year-old female with a history of relapsing polychondritis, RA, asthma and Celiac disease presented with a chief complaint of recurrent sinusitis. She suffered from recurrent pharyngitis and sinusitis since childhood, and has undergone 3 sinus surgeries. Her presentation of RP began with episodes of paroxysmal ear pain at age 10. She subsequently developed widespread arthralgias. Lab evaluation was significant for detectable CCP antibody. ESR, CRP, Interleukin-1 beta, interleukin-6 and TNF levels were all markedly elevated. She was found to have selective IgA deficiency. However, she had an adequate antibody response after vaccination with pneumovax. Celiac was diagnosed based on biopsy. Initial treatment with NSAIDs, hydroxycholoroquine and colchine failed to alleviate her symptoms. Treatment with TNF-inhibitors improved her arthralgias, but caused worsening injection site reactions which led to their discontinuation. She is now beginning treatment with abatacept.

Conclusion: RP is an often misdiagnosed, serious medical condition affecting multiple organ systems. Allergists/immunologists should keep a high index of suspicion for this entity, especially when caring for patients with comorbidities commonly associated with RP. Whereas RP is often associated with autoimmunity and IBD, this case is unique as Celiac disease and IgA deficiency are uncommon comorbidities.

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KOUNIS SYNDROME PRESENTING IN A PEDIATRIC PATIENT IN THE SETTING OF MAST CELL ACTIVATION DISORDER

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Introduction: Kounis syndrome is a rare condition that is classically associated with vasospastic angina or acute myocardial infarction in the setting of mast cell activation and the release of inflammatory mediators. Mast cell activation disorder is characterized by an excessive inflammatory reaction of mast cell mediator release resulting in chronic symptoms and anaphylaxis. To our knowledge, this is the first documented case of Kounis syndrome in a pediatric patient with mast cell activation disorder.

Methods: Case presentation.

Results: A 17 year old female with history of Ehlers Danlos syndrome, Chiari type I malformation with craniocervical instability requiring multiple spinal surgeries, mast cell activation disorder on cromolyn, montelukast and hydroxyzine presented in cardiogenic shock several days after spinal fusion surgery. A postoperative transfusion resulted in progressive tongue swelling and respiratory distress requiring an emergent airway. She was found to have elevated troponins and electrocardiogram changes consistent with acute myocardial infarction. Echocardiogram revealed a severely diminished left ventricular ejection fraction of 22%. A cardiac MRI was done which showed that the cardiac insult was not ischemic in nature. She slowly recovered after being treated with an aggressive corticosteroid taper and several week course of omalizumab. Her cardiac function returned to normal.

Conclusion: This patient presented with a rare complication of mast cell activation in the form of significant cardiac injury. Detection of Kounis syndrome is vital for both acute management and long term control in these patients.

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HYPOGAMMAGLOBULINEMIA AS PRESENTING SIGN OF NEPHROTIC SYNDROME

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Rationale: Secondary hypogammaglobulinemia has been described. We report a case of severe rapid-onset hypogammaglobulinemia presenting as adult onset nephrotic syndrome.

Background: Hypogammaglobulinemia has been reported in pediatric steroid sensitive nephrotic syndrome[1]. Similar findings in adult onset nephrotic syndrome in the A&I literature have not been found.

Case Presentation: A 59-year-old white hypertensive male ex-smoker presented 1/14 with severe persistent asthma and history of metastatic prostate cancer on Lupron therapy in remission. FEV_1 was 2.68 L. Over the next two years, he had asthma flares requiring prednisone. In fall 2015, echo and cardiac stress tests were normal. Labs showed IgG 770; IgA 111; IgM 66; IgE 17; normal ESR, CRP, albumin, creatinine, and globulin. TEC was 456. He started Mepolizumab. By 12/16, TEC was 0. He acutely developed dyspnea, anasarca, and bilateral pleural effusions 3/17. FEV_1 was 1.68 L, BNP was elevated, albumin and globulin were decreased, ANCA and ANA were negative. Dyspnea did not respond to anti-asthmatic or diuretic therapy. Cardiac catheterization revealed normal coronaries with high output failure. In 4/17, IgG was 200, IgA 81, IgM 59, urine showed 3+ proteinuria, negative sediment, urinary polyclonal elevations in kappa and lambda light chains, albumin 2.6, creatinine 1.4, globulin 2.0, and 24-hour urine protein 2.85 g. Kidney biopsy revealed focal segmental glomerulonephritis (FSGN). Causes of FSGN were excluded by labs and imaging. Steroids and IVIg were initiated.

Discussion: This is a rare case of hypogammaglobulinemia presenting with nephrotic syndrome. Dyspnea was secondary, not to asthma, but fluid overload from acute nephrotic syndrome from idiopathic FSGN.