Immunoglobulin A Vasculitis Presenting with Gastrointestinal Symptoms: A Case Presentation

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A 69 year old Caucasian male presented to the hospital for his first dermatology clinic visit at the age of 60 days, history of abdominal pain, vomiting, and bloody diarrhea. Additionally, it was noted upon admission that he had a rash on his bilateral lower extremities extending proximal to his abdomen. According to his living facility, the rash had started 4 days prior to admission. He denied any genitourinary symptoms or symptoms of the rest of his review of systems was negative. His past medical history included hypertension, hyperlipidemia, gastroesophageal reflux disease, colitis, insomnia and depression for which he was being treated with metoprolol, atorvastatin, omeprazole, probiotics, zolpidem, and citalopram. Prior surgeries included an appendectomy and left inguinal hernia repair. Physical exam revealed multiple purpuric macules on his feet, legs, hips, and abdomen. Prior laboratory findings included an elevated sedimentation rate (162), positive fecal occult blood test, and abnormal urinalysis with 2+ blood and 1+ protein. Abdominal imaging revealed no acute or chronic pathology to demonstrate significant findings. Skin biopsies for routine histopathology and direct immunofluorescence were performed at time of clinical presentation due to suspected vasculitic vascular deposition of IgA. The patient was treated with intravenous pulses of solumedrol every 8 hours that was gradually tapered with improvement.

While the cause of IgA vasculitis is unknown, several theories exist. It is clear that IgA plays a central role, with deposition of immune complexes between antigens and IgA in the skin, gut and kidneys. It has been proposed that antigen presented by mucosa-associated immune system leads to IgA synthesis3 and immunoglobulin deposition in vessel walls results in complement activation and vascular damage22. An infectious cause could also be considered, especially in children who tend to present with IgA vasculitis 1-2 weeks following an upper respiratory infection. Additionally, the fall/winter peak and self-limited course are also congruent with the infectious theory. However, no consistently specific causative agent has been identified and various cases have been linked to a wide array of pathogens, including bacteria (such as group A β-hemolytic streptococci), viruses, and parasites23. Pharmacologic agents, such as antibiotics, antitumor necrosis factors and chemotherapeutic agents, as well as vaccines have also been implicated. In adults, cases have been linked to cancer, possibly reflecting a tumor induced immune response. Genetics may also play a role. Certain human leukocyte antigen alleles, such as HLA-DRB1*04, HLA-DRB1*11, HLA-B8 and HLA-A11, have been found to be associated with increased risk of IgA vasculitis22. While another, HLA-DRB1*08, was found to be protective24. A link has also been established between IgA vasculitis and familial Mediterranean fever gene mutations. Workup of IgA vasculitis should include a thorough history including queries regarding recent illnesses, exposures to medications, vaccines or chemicals, history of autoimmune disease or malignancy, and review of systems. In addition to history and physical, biopsies for BHE and direct immunofluorescence, and laboratory analyses should be ordered. Labs should include, at minimum, complete blood count with differential, erythrocyte sedimentation rate, urinalysis, serum creatinine, and urine protein. Based on history and clinical suspicion, one may also consider C-reactive protein, serum and urine protein electrophoresis, cryoglobulins, liver function tests, fecal occult blood test, urinalysis, hepatitis panel, anti-neutrophil cytoplasmic antibodies, and complements 3 and 4. In patients with gastrointestinal symptoms, imaging studies (including abdomino-scan and CT) as well as esophagogastroduodenoscopy and colonoscopy are appropriate.

Since the majority of symptoms are self-limited, treatment is mainly supportive. Corticosteroids, while controversial, should be considered for severe systemic manifestations24. Steroids may improve early GI symptoms by decreasing intestinal wall edema and also prevent complications such as bleeding and intussusceptions. However, they do not prevent GI recurrences. Steroids or cytocytic medications may also be helpful for preventing renal sequelae25. yet exact guidelines in management remain undefined. Other anecdotal reports describe beneficial effects with plasmapheresis26, monoclonal antibody27 and Plasmapheresis: a review article. Southern Med J 2007; 100:821-824.


