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A CASE SERIES ON ADULT-ONSET IGA VASCULITIS

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ABSTRACT

Immunoglobulin IgA vasculitis, also known as Henoch-Schönlein purpura, is the most prevalent type of systemic vasculitis in children and typically develops between the ages of 3 and 15. The reports of its occurrence in adults is rare. The low prevalence in adults is probably due to misdiagnosis or improper diagnosis. In order to properly manage patients and prevent its related complications, physicians should be able to correctly identify IgA vasculitis in addition to the pediatricians. There is still some research to be done on the management of adult-onset IgA vasculitis with multi organ involvement. Here, we report a case series of three adult-onset IgA vasculitis cases that were triggered by different elements.

INTRODUCTION:

Immunoglobulin A vasculitis (IgAV) is an immune complex mediated small vessel leukocytoclastic vasculitis that is common in children but rare in adults. It is characterized by the presence of IgA deposits in the small vessels of skin, gastrointestinal system, joints and kidneys [1]. Approximately two thirds of patients have gastrointestinal (GI) complaints like nausea, vomiting, oedema and abdominal discomfort. Ischemia and oedema are the causes of these symptoms, and arthralgia occasionally results in infarction, intussusception, or intestinal perforation.[2] While it is often benign in childhood, in adults it has more severe disease progression and need more aggressive treatments. Due to its rarity, the understanding of its pathogenesis, natural history, and disease course in adults is limited. [3] Regarding the use of immunosuppressive drugs, current therapy recommendations are still nebulous and debatable. There is also no consensus in the management of IgAV in patients with severe multi-organ involvement. [4] At the moment, the overall strategy for the initial workup and management of IgA vasculitis is dependent on comorbidities, related symptoms and experience of the clinician managing the patient at the time of diagnosis. The management often involves complex and long term systemic immunosuppression.[5] Randomized controlled studies that could inform therapy guidelines are difficult to conduct due to the low incidence. So, we present three cases of IgA vasculitis in adults who showed up with different symptoms, which are triggered by different elements in each.

Case Report:

Case 1:

A 28-year-old male patient came with symptoms of generalized swelling over the body for 2 days. It was sudden in onset, which progressed from face to the rest of the body. No other symptoms were noted. There was no relevant past history. On physical examination, there was bilateral pitting edema, along with anasarca [Image 1]. Local examination of the peripheries revealed two ulcers of 2x2 cm- with one in the medial aspect of left ankle, and one behind the medial malleolus. There was blackish discoloration of slough, but no pus or warmth on the ulcer [Image 2]. The differential diagnosis was angioedema under evaluation due to nephrotic syndrome or nephritic syndrome. The patient's investigations showed negative ANA, serology and other markers like C3, C4, C-ANCA, IGE etc., Urine analysis showed a spot urine PCR of 0.35. Upon the dermatologist's suggestion, a wedge biopsy was taken from the left lower leg ulcer site. Meanwhile, the patient was treated with IV hydrocortisone, as the swelling started decreasing. On probing further history, patient revealed red ant bite in the temple 4 days prior to the onset of illness. Rheumatologist opined that vasculitis could be the diagnosis. Serum IgA levels and the markers of inflammation were investigated.

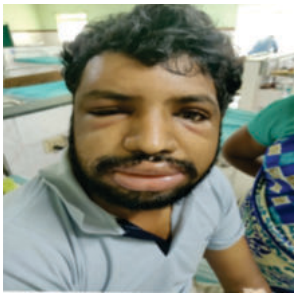

Image 1

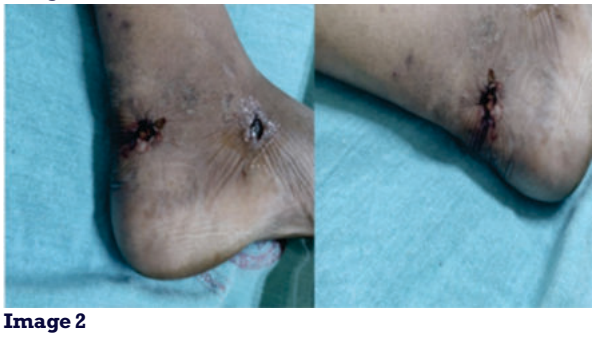

Image 2

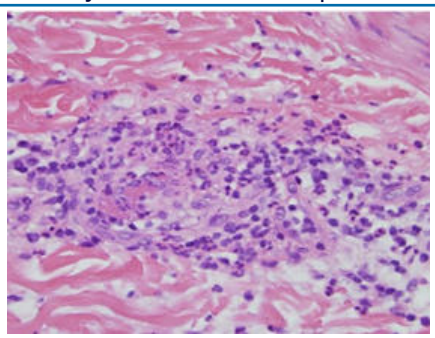

Image 3

Three days after the admission, the patient developed non-blanchable purpuric rash on his hands and body. Histopathology of the biopsy from the ulcer showed neutrophilic infiltrations and congested blood vessels. There were foci of necrotic material and karyorrhectic debris in its wall [Image 3]. IgA levels of 512.7 mg/dl has confirmed the diagnosis of leukocytoclastic vasculitis, direct immunofluore scence was done and showed deposition IgA and confirmed the diagnosis. The patient was kept on glucocorticoid therapy until the symptoms subsided then tapered and stopped, and the prognosis of the patient was extremely good, with recovery made in few days.

Case 2:

The 27-year-old married female patient attended the tertiary care hospital with complaint of abdominal pain and vomiting for the past 4 days. There was no other relevant history. After admitting, she had painful, bilious vomiting, and developed mild ascites. She was kept on intravenous fluids and symptomatic treatment, and the investigations were carried out, such as CT scan, complete blood picture, liver and renal function tests. GI endoscopy showed a possibility of hiatus hernia or GI vasculitis. She developed reddish purple spots over buttocks, thighs and legs. Skin biopsy from a lesion from left forearm was sent, that showed features of leukocytoclastic vasculitis [Image 4] and direct immunofluorescence was done and showed deposition of IgA and confirmed the diagnosis. Her complement (C3, C4), P-ANCA, L-ANCA came negative. CT abdomen suggested the presence of duodenal enteritis with diffuse thickening of duodenum, which might be secondary to Henoch-schonlein purpura. The patient was kept on glucocorticoid therapy until the symptoms subsided then tapered and stopped, and the prognosis of the patient was extremely good, with recovery made in few days.

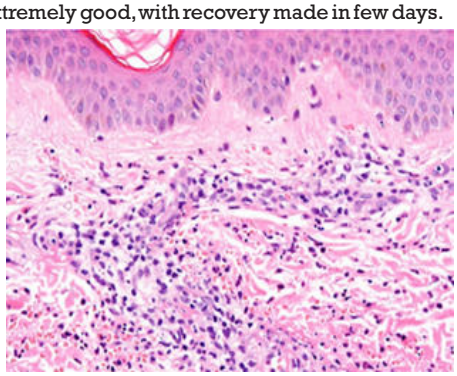

Image 4

Case 3:

A case of married female, 31 years old, has presented with cutaneous vasculitis of both lower limbs to General medicine department. She was found to have taken siddha medicine, which triggered the rash. The skin rashes eventually increased in number and spread to the rest of her body. There were tender purpuric papules with central crusting [Image 5]. She developed pain in joints, muscle spasm, abdominal pain, and vomiting withing the next two days of admission.

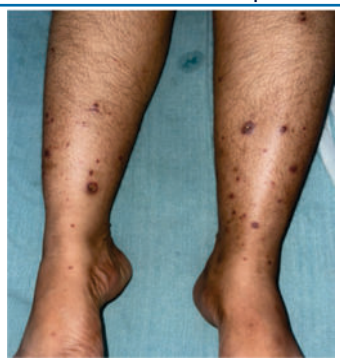

Image 5

Ultrasound abdomen revealed splenomegaly, hepatomegaly, and umbilical hernia. Urine culture showed presence of bacteria, albumin, RBCs. There was elevated C-reactive protein. The C3 value was very high, while ANCA values were in normal limit. Biopsy of the skin lesion showed small vessel vasculitis associated with epidermal neutrophilia and spongiosis [Image 6]. Immunofluorescence showed granular florescence of IgM, IgA, C3c and fibrinogen seen in the papillary dermal vessels. The patient recovered spontaneously in few weeks, with glucocorticoid therapy.

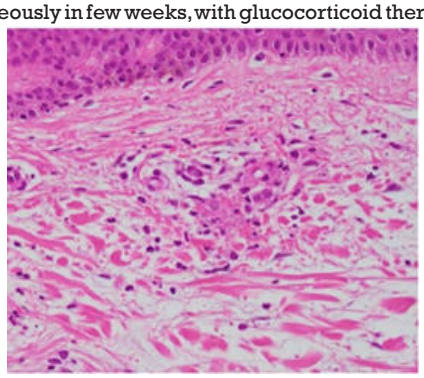

Image 6

DISCUSSION:

The yearly incidence of IgA vasculitis in adults ranges from 0.8 to 2.2 per 100,000 people. Males are five times more likely to contract the illness than females, and it is known to have seen more in those with obesity, diabetes, and hypertension [6]. These pre-existing comorbidities were known to cause early deaths in those with IgA vasculitis. IgA1-dominant immune complexes are thought to be a major factor in the etiology of the disease. IgA vasculitis can be presented differently in childhood with monophasic presentation than the disease's clinical symptoms as an adult. The common spectrum of illnesses includes skinny purpura (61%) arthralgia (61%), acute enteritis (48-53%), and glomerulonephritis (45-85%) [7]. All the three patients have shown visible purpura in our case series. Severity predictors could not be assessed in these cases as well. Abdominal pain, discomfort and vomiting were seen in the two patients, and it was mostly due to bowel ischemia and edema in both.

Renal complications are one of the primary variables affecting the long-term morbidity and fatality of IgAV. 11% of patients with kidney involvement will eventually acquire end-stage renal disease. [8] One of our patients had renal involvement, which was resolved with the standard treatment. There are many unanswered questions regarding the diagnosis and prognosis of severe organ-threatening IgAV in adults. While mild to intermediate visceral organ manifestations of IgAV can usually be treated symptomatically, severe visceral organ manifestations require complex and long-term immunosuppression. [9]

IgAV is a self-limiting form of systemic vasculitis, and most

patients heal from it on their own. As a result, supportive care, which in the vast majority of cases also includes sufficient hydration, rest, and pain relief through symptoms, is the main focus of management. There hasn't been a report evaluating treatment options for adult-onset IgAV, and the data from studies involving children casts doubt on the use of glucocorticoids or immunosuppressants. [10] Oshikata et al. reported a case of catastrophic IgAV with non-occlusive mesenteric ischemia that resulted in death despite aggressive immunosuppression, including IV steroids and cyclophosphamide. [11] Glucocorticoid therapy is said to speed up the resolution of arthritis and abdominal pain, but it doesn't seem to stop the illness from coming back. Therefore, routine glucocorticoid therapy is not advised for IgAV patients to treat symptoms or avoid renal or gastrointestinal complications. Only those patients with nephrotic syndrome whose symptoms are severe enough to conflict with their ability to eat, walk, or perform activities of daily living and/or necessitate hospitalization may receive recommendations for glucocorticoid therapy.[12]

CONCLUSION:

We come to the conclusion that physicians should consider IgAV in the differential diagnosis of leg purpura in both infants and adults. We discuss our experiences using intensive immunosuppression to treat serious systemic IgAV. To induce and sustain remission in our patients with a high burden of disease, aggressive treatments were necessary. The absence of treatment guidelines makes it more difficult to make choices about treatments and to predict their results and prognosis. Balancing disease prevention and treatment-related complications calls for a measured and thoughtful strategy. It will be very beneficial to conduct more research on the function of biologics and other cutting-edge drugs in severe IgAV. Early use of biologics may lessen the load of toxicities associated with treatment.

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