Case report

A Rare Case of Adult Onset IgA Vasculitis

Abstract:

Background: IgA vasculitis, formerly known as Henoch – Schoenlein Purpura (HSP), is vasculitis of small vessels, which is a rare and life threatening condition in adults. It is a type 3 hypersensitivity reaction which can affect kidney, joints, skin and intestine. It is a rare presentation in adults and more severe than its pediatric counterpart.

Case Presentation: A 26 year old male with no comorbidities presented with complaints of non-resolving macular rashes on bilateral lower limbs and abdominal pain associated with fever and blood stained loose stools. Upper GI Endoscopy showed features of duodenitis and skin biopsy confirmed the diagnosis of IgA Vasculitis. Patient was started on steroids and was discharged as he improved symptomatically.

Conclusion: Clinical suspicion for adult onset IgA vasculitis should increase in the clinical practice. Early detection of the disease and early initiation of appropriate treatment help in improvement of prognosis of complications associated with IgA vasculitis.

Keywords: IgA vasculitis, prognosis, bilateral lower limbs, small vessel vasculitis

Introduction:

IgA vasculitis is immune-mediated small vessel vasculitis in which deposition of IgA-dominant immune complexes leads to pathological involvement of the skin, kidneys, gastrointestinal tract, and joints. It is the most common form of childhood vasculitis, accounting for 45% of pediatric vasculitis with an incidence of 20 per 100,000 per year in children less than 17 years of age. [1-5] The disease is rare in adults, with an annual incidence of 0.1 to 1.8 per 100,000 individuals. It is usually benign in children, but adult-onset IgA vasculitis has a severe course of the disease requiring aggressive management, and it has a bad prognosis when associated with renal involvement. Some of the rare cases of adult onset have been published.[12-15,18]

About two-thirds of the adult cases present with GI symptoms, e.g., nausea, vomiting, abdominal pain, and bloody stools. While any part of the gastrointestinal tract may be involved, the small bowel, especially the duodenum, is most commonly affected.

Case scenario:

A 26-year-old male with no known comorbid conditions presented to the emergency department with complaints of fever, blood-tinged loose stools, and diffuse pain in the abdomen for 15 days. He subsequently developed macular rashes on bilateral lower limbs. He presented to our emergency department with worsening symptoms.

On initial evaluation, his vitals were unremarkable. Abdominal examination revealed tenderness in the epigastric region, otherwise normal. On examination, non-palpable, non-blanching, macular rashes suggestive of purpura on bilateral lower limbs and abdomen were found.

Initial investigations revealed a serum lactate of 3.1mmol/L. USG abdomen showed mildly edematous and hyperemic wall of the 1st part of duodenum.

The patient was treated symptomatically, and he was admitted to the observational ward with a probable diagnosis of HSP.

An UGI endoscopy with duodenal biopsy was done, which showed duodenal mucosa with focal crypt villous broadening. Lamina propria showed mild infiltration of lymphocytes, plasma cells, and occasional eosinophils suggestive of duodenitis.

ANA profile, c-ANCA, p-ANCA, GBM antibodies negative.

Immunology and dermatology opinions were sought. He was treated symptomatically with Inj. Ciprofloxacin and Tab. Fenofibrate for 5 days. Initially the patient improved symptomatically but subsequently had worsening rashes spreading to his upper limbs and back. Symptoms subsided with steroids.

Skin biopsy and immunofluorescence showed granular positivity for IgA in blood vessels and is negative for IgG, IgM, and C3.

Table 1 : Pathological test result

Hb	14.7 gm/dl
TLC	14450 10^9/L
N/L/E/M/B	75%/17%/0.4%/7%/0.1%
Platelets	248000 10^9/L
PT	12.1
APTT	32.4
Peripheral Smear	Normal picture
Urine microscopy	Normal
Blood culture	No Growth
Stool culture	No growth

Stool microscopy	No WBC/RBC/Ova/Mucous

Discussion and Conclusion

Adult-onset IgA vasculitis is a rare disorder with an incidence of 0.8–5.1 per 100,000 individuals, with increased frequency in the fifth and sixth decades of life. [7] Several retrospective studies show 20 to 30% of patients with IgA Vasculitis were adults. In these cohorts, adults had significantly worse kidney outcomes compared with children.

IgA-antibody immune complexes, which are caused by antigenic exposure from an infection or medication, get deposited in the small vessels (usually capillaries) of the skin, joints, kidneys, and gastrointestinal tract. This causes increased production of prostaglandins. If these immune complexes get deposited in the intestinal wall, they may cause gastrointestinal bleeding. If the deposition occurs in kidneys, it may cause crescentric glomerulonephritis. Immune complex deposits in the skin cause palpable purpura. [9]

The HSP tetrad consists of purpura, arthritis, nephritis, and abdominal pain. The incidence of gastrointestinal involvement was 80% in an Indian study of adult-onset HSP~~6~~.[6] It affects the small intestines with preference for the second portion of the deuodenum. [4] If GI is involved, patients may have nausea and vomiting that worsens after meals. Patients may present with malena, hemorrhagia, or acute abdominal pain. Because of their similar presentations, IgA vasculitis should be differentiated from inflammatory bowel disease. Endoscopic evaluation aids in quick and accurate diagnosis and differentiates between the two disease conditions.

Skin is involved in all patients with IgA vasculitis. The rash associated with the disease is non-pruritic, and it is characterized by palpable purpura and petechiae that most commonly affect the buttocks and lower extremities, particularly the extensor surfaces. Skin involvement is present in all patients with IgA vasculitis.

Renal manifestations include hemorrhagia, proteinuria, nephrotic syndrome, nephritic syndrome, and renal failure. The most common renal manifestation is microscopic hemorrhage. Severe proteinuria may present as nephrotic syndrome, and patients with persistent proteinuria are at high risk of developing progressive glomerulonephritis. [10]

Patients often present with painful swollen joints that most commonly involve the knees, ankles, hands, and feet. The arthritis is typically transient and non-destructive. According to EULAR criteria, a patient was classified as HSP in the presence of purpura or petechiae (mandatory) with lower limb predominance plus one of four criteria: 1) abdominal pain; 2) histopathology (immunoglobulin A (IgA)); 3) arthritis or arthralgia; and 4) renal involvement. Our patient presented with palpable purpura, worsening abdominal pain, which fulfilled the diagnostic criteria for HSP. [11]

IgA Vasculitis, or Hench Schoenlein Purpura, is a self-limiting disease, and the treatment is mainly supportive[16]. The use of non-steroidal anti-inflammatory drugs (NSAIDs) is effective in relieving arthritic pain and abdominal pain. Glucocorticoids have been successfully used to treat the disease by reducing abdominal pain[16]. However, glucocorticoids have not been shown to decrease the renal complications associated with the disease.[17]

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