



ADULT-ONSET IMMUNOGLOBULIN A VASCULITIS

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ABSTRACT Immunoglobulin A vasculitis (IgAV), formerly Henoch–Schönlein purpura vasculitis, is a vasculitis commonly seen in children and only rarely described in adult patients. IgAV can present as arthralgia, rash, discolored urine, acute kidney injury, and gastrointestinal symptoms. We present a case of a 60-year-old man who presented with red colour skin lesion on bilateral lower extremity, upper extremity and on buttocks since 15 days followed by worsening abdominal pain. Computed tomography suggestive of ileocolitis. Upper GI scopy and colonoscopy revealed Duodeno-jejunal, Terminal ileal ulcerations and in GI biopsy and skin biopsy confirmed a diagnosis of IgA vasculitis.

KEYWORDS :

INTRODUCTION

Henoch-Schönlein (IgA vasculitis) is a small-vessel vasculitis characterized by palpable purpura (most commonly distributed over the buttocks and lower extremities), arthralgias, gastrointestinal signs and symptoms and glomerulonephritis. Henoch-Schönlein (IgA vasculitis) is usually seen in children; most Patients range in age from 4 to 7 years; however, the disease may also be seen in infants and adults. It is not a rare disease; in one series it accounted for between 5 and 24 admission per year at a pediatric hospital. The male-to-female ratio is 1.5:1. A seasonal variation with a peak incidence in spring has been noted.

CASE HISTORY

A 60-year-old white man with a medical history of hypertension admitted with chief complain of red colour skin lesion on bilateral lower extremity, upper extremity (figure 1) and on buttocks since 15 days followed by worsening abdominal pain and severe joint pain in both elbow and wrist. He started bleeding per rectum since 2 days after admission. His abdominal examination revealed a soft, nontender, nondistended abdomen with bowel sounds present. The dermatological examination revealed a nonpalpable purpura on multiple sites.

Initial laboratory evaluation revealed a leukocytosis of $19 \times 10^9/L$ and serum creatinine within normal range. Other investigation like C3, C4 was within normal limit, ANA was negative, viral markers negative, stool for occult blood positive and in urine routine micro examination albumin was present. Inflammatory markers ESR was high (60) and CRP also positive.

USG ABDOMEN suggestive of Short segmental circumferential wall thickening/mucosal oedema involving 4th part of duodenum and proximal jejunal loop with surrounding few hypoechoic subcentric lymph nodes. Long segmental edematous wall thickening involving terminal ileum, IC junction, caecum & proximal ascending colon – ileocolitis. Computed tomography with intravenous contrast of the abdomen revealed terminal ileitis. In UPPER GI SCOPY Portal gastropathy and Duodeno-jejunal ulceration seen (figure 2 & 3). In COLONOSCOPY Terminal ileal ulcerations. Colopathy, likely PHTN related (figure 4). Biopsy from duodenum & terminal ileum suggestive of IgA Vasculitis. Skin Biopsy from left lower leg on shin of tibia findings are consistent with leukocytoclastic vasculitis.

He was given 1,000 mg methylprednisolone and then 500 mg methylprednisolone the 5 days follow by tapering dose according to 1g/kg/day and started tablet azathioprine 50mg in once a day dose. Patient is improved after this treatment and discharged after 8 days.



Figure 1

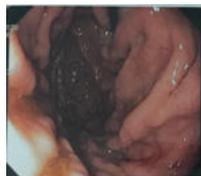


Figure 2

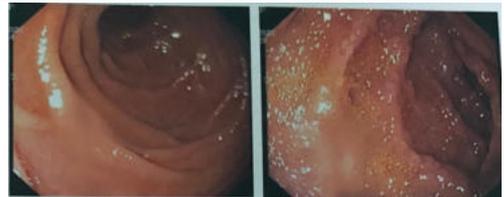


Figure 3

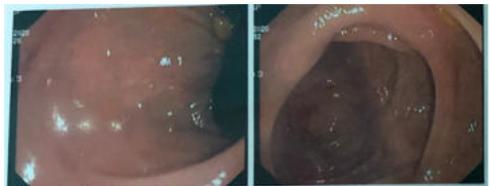


Figure 4

CONCLUSION

IgA vasculitis is the most common vasculitis to affect the upper GI tract with one study finding it to account for 56.8% of all cases. Although the duodenum is the most common location affected by IgA vasculitis, any portion of the GI tract may be affected, including less commonly the terminal ileum. Because of their similar presentations, endoscopic evaluation can aid in making a diagnosis because IgA vasculitis will classically have endoscopic findings of diffuse mucosal erythema, petechiae, hemorrhagic erosions, and longitudinal ulcers. A biopsy is often needed to help establish a distinction between IBD and IgA vasculitis, although there has been at least one case report of IgA vasculitis with endoscopic biopsies consistent with IBD. Although biopsy of the leukocytoclastic rash which accompanies IgA vasculitis will distinguish it from IBD, up to 14% of the cases will present with abdominal symptoms before the manifestation of a rash. It is important to make an accurate diagnosis between IgA vasculitis and IBD because the long-term treatment is different for the diagnoses. One study found that the most commonly used treatment in IgA vasculitis being renin-angiotensin-aldosterone system blockers and corticosteroids. Although many medications can be used for the long-term treatment of IBD, that same study found that azathioprine was the only medication also used in the treatment of IgA vasculitis and was only found to be used in 6.1% of all adults with IgA vasculitis.

ACKNOWLEDGEMENT

I would like to thank all three of my guide and mentor for their full support throughout the process.

1. DR.M.N.SAIYAD (Head of Unit, General Medicine department, SCL General Hospital, Ahmedabad, Gujarat, India.
2. DR.SHEETAL D. VORA (Associate Professor, General Medicine department, SCL General Hospital, Ahmedabad, Gujarat, India.
3. DR.SNEHA PATEL (Assistant Professor, General Medicine department, SCL General Hospital, Ahmedabad, Gujarat, India.

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