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IgA Vasculitis Mimicking Obstructed Umbilical Hernia and Presenting as Gastrosurgical Emergency

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Abstract

IgA vasculitis (previously known as Henoch-schonlein purpura) is a systemic vasculitis which is more common in children than in adults. The classical triad clinically suggests the diagnosis includes purpuric rash, arthritis and abdominal pain. We report a case of a forty-seven-year-old male presented with an acute abdomen to a gastro-surgeon, and on exploratory laparotomy, multiple petechial lesions were found on the entire intestinal walls. Multiple petechiae were present over both the legs and hands as well, which prompted a dermatological consult, and the diagnosis of IgA vasculitis was confirmed with histopathological and immunofluorescence evaluation of the skin lesions.

Key words: Abdominal pain; Acute renal failure; Henoch Schonlein purpura; IgA vasculitis; Petechiae

Introduction

IgA vasculitis / Henoch Schonlein Purpura (HSP) is a common type of vasculitis in which the small blood vessels are involved with immune complex (IgA and complement 3) deposition on arterioles, capillaries and venules. It is predominantly a disease of childhood with rare occurrences in adults.[1] Certain triggering factors have been identified, which include medications, environmental factors and preceding infections with agents like group A streptococcus, parvovirus B19, Bartonella henselae, H.parainfluenzae, Coxsackie virus, and methicillin-resistant staphylococcus. After an antigenic exposure from an infection or medication, IgA-antibody immune complexes are formed and get deposited in the small vessels (usually capillaries) of the gastrointestinal tract, dermis, joints and kidneys, producing an influx of inflammatory mediators like prostaglandins. Complement C3 receptor lymphocytes may bind to these immune complexes and deposit in the vessel walls, aggravating the inflammatory response. Hence, systemic manifestations are seen commonly in HSP, along with cutaneous manifestations of palpable purpura and petechiae. Immune complex deposition in various organs may lead to gastrointestinal haemorrhage, mild proliferative or severe crescentic glomerulonephritis and arthritis.

Case Report

A forty-seven-year-old male having a long-standing umbilical hernia presented to the gastro-surgeon with complaints suggestive of an acute abdomen since 1 day. On taking history, he revealed 2-3 episodes of fever seven days back, for which he had taken multiple antipyretic drugs. The surgeon noted some reddish lesions over the legs, which the patient had also noticed in the last few days, along with swelling over both feet. Considering the possibility of an obstructed umbilical hernia (acute abdominal pain and vomiting with tender umbilical hernia), the patient was evaluated with ultrasonography, which revealed oedema of the

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bowel wall, including the loops in the herniated sac. The patient underwent urgent exploratory laparotomy, where the surgeon found multiple petechiae over the entire intestinal walls with no signs of obstruction or intussusception. Hernioplasty was done using mesh to cover the wall defect. (Fig 1) The conundrum of mucosal petechiae and cutaneous lesions prompted a dermatologist's opinion. Detailed cutaneous evaluation revealed multiple petechiae over both legs, periumbilical area, buttocks and hands, which were progressive over the past five to seven days.(Fig 2)

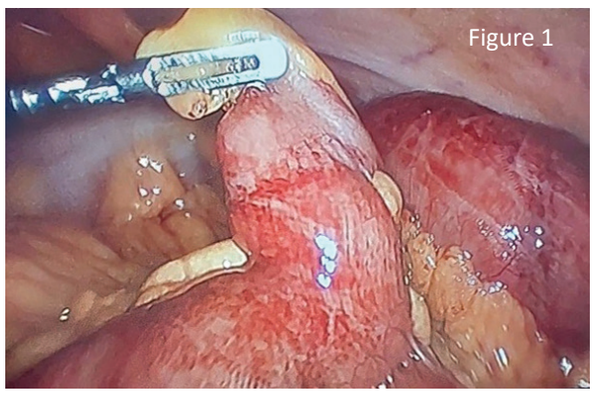

Figure 1: Multiple petechiae on gastrointestinal mucosa

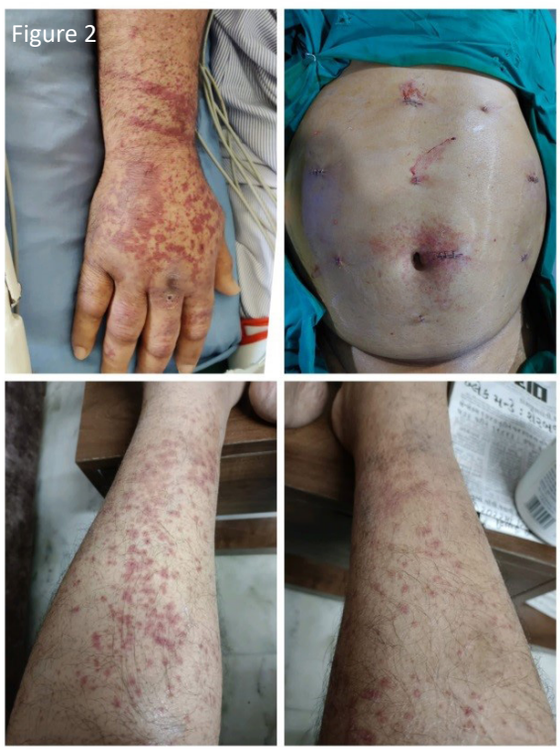

Figure 2: Multiple petechiae over hand, periumblical area and both legs

The patient also had decreased urine output, which was secondary to acute kidney injury, and the patient was put on haemodialysis. Haematological investigations showed low haemoglobin, raised c-reactive protein (CRP) and altered renal profile. The coagulation profile was normal. Urine examination showed multiple red blood cells. Two biopsy specimens were sent from the skin lesions for histopathology and direct immunofluorescence (DIF) examination. Histopathology showed vasculitis in small-sized vessels with mural neutrophilic infiltrate and fibrinoid necrosis along with extravasated red blood cells in the dermis. DIF showed IgA and IgG staining in the dermal vessel wall (Fig 3).

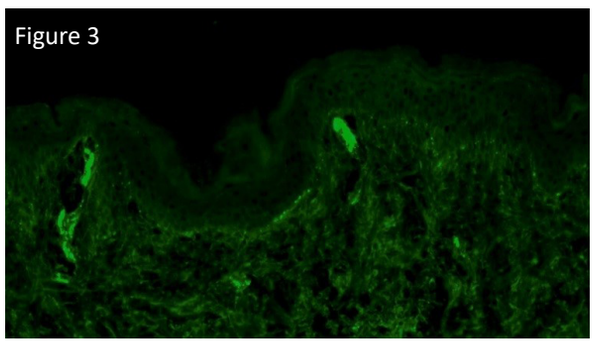

Figure 3: IgA deposition in vessel walls on DIF

In view of the classical constellation of symptoms and signs involving the skin, kidneys and gastrointestinal tract, the final diagnosis of IgA vasculitis was made. In view of severe renal involvement, the patient was started on systemic steroids, and the patient improved over a period of 20 days. The renal function returned to normal, and the patient did not have any further complaints on follow-up for 1 year.

Discussion

A triad of purpuric rash, abdominal pain and arthritis characterises IgA vasculitis. The incidence of the HSP in adults is 3.4 to 14.3 cases per million population.[1] The classic symptoms include a purpuric rash, which appears on the legs and progresses to the buttocks. The rash may also be seen on the arms, face and trunk. Other atypical lesions which can be seen are urticaria, vesicles, bullae, targetoid lesions, and foci of necrosis. Abdominal pain occurs in around 56% of the patients, and 15% of the patients present with abdominal pain. Joint involvement presents as non-erosive arthritis involving ankles, knees and elbows and hence does not cause permanent deformity. Approximately 40% of patients will have renal involvement at some point of the diseases course, which manifests as microscopic haematuria. When kidneys are involved, proteinuria occurs in most of the patients, and only a minor proportion progresses to nephrotic syndrome, while only 1% of patients progresses to develop chronic kidney disease.[2] HSP is associated with a higher frequency of systemic involvement in adults, and complete recovery has been observed in the majority of the cases.[3]

A multi-speciality approach including a dermatologist, nephrologist and gastro-surgeon is required since the patient can present with purpuric rash to a dermatologist or with features of kidney involvement to a nephrologist or with symptoms of acute abdomen to a gastro-surgeon. A criterion has been proposed for the diagnosis (Table 1) [4]

Table 1: Diagnostic Criteria for Henoch Schonlein Purpura as proposed by EULAR/PRINTO/PRES (2010)

Criterion	Description
Mandatory Criteria	Purpura and petechiae with lower limb predominance
Minimum one out of four criteria	Acute Onset diffuse abdominal pain
	Histopathology showing leucocytoclastic vasculitis with predominant IgA deposit or proliferative glomerulonephritis with predominant IgA deposits.
	Arthritis or arthralgia of acute onset
	Renal Involvement showing proteinuria >0.3 g/24 h or >30 mmol/mg of urine albumin/creatinine ratio on a spot morning sample, Haematuria or red blood cell casts: >5 red blood cells/high power field or red blood cells casts in the urinary sediment or ≥2+ on dipstick

EULAR: European League Against Rheumatism, PRES: Paediatric Rheumatology European Society, PRINTO: Paediatric Rheumatology International Trials Organisation.

In our case, though the patient correctly presented to a gastro-surgeon for acute abdomen, an invasive procedure may have been deferred or completely avoided with earlier involvement of a dermatologist for cutaneous lesions of palpable purpura and petechiae in the setting of acute kidney injury.

Conclusion

Our report highlights the need for gastro-surgeon, nephrologists and dermatologists to be alert to gastrointestinal manifestation with multiple purpuric skin lesions and involvement of the kidney, especially when lesions are above the waist line.

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