INTERNAL MEDICINE

Henoch-Schönlein Purpura with Upper Gastrointestinal Hemorrhage in an Adult

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ABSTRACT

Henoch-Schönlein purpura (HSP) is a small vessel vasculitis characterized by palpable purpura, arthralgia, gastrointestinal (GI) signs and symptoms and glomerulonephritis. It is disease of children. But when it effects adults it can be more severe and outcome may be relatively poor. Both significant upper GI bleed and scrotal involvement were not reported in adults many a times in the literature. Here we present adult patient with HSP who presented with GI hemorrhage and scrotal swelling with good clinical outcome.

Keywords: Henoch-Schönlein purpura, acute abdomen, gastrointestinal hemorrhage, scrotal swelling, adult

'enoch-Schönlein purpura (HSP) is an immunoglobulin A vasculitis, most commonly affecting the children of age group 3-15. The annual incidence is 10-20/1,00,000 children aged below 17 years. It is rare in adults, approximately 10% HSP cases occur in adults and disease can be more severe and may not be self-limiting as in children.² HSP is a disease of unknown etiology, thought to be triggered by various infections and immunizations. It is characterized by a tetrad of symptoms, a palpable purpura without coagulopathy or thrombocytopenia, arthralgia or arthritis, abdominal pain or renal disease. There is no specific test for HSP; diagnosis is clinical. Diagnostic dilemma may occur if the disease presents incomplete. In such situations, biopsy of affected organ is warranted, which will reveal predominantly immunoglobulin A deposits.

CASE REPORT

A 55-year-old male patient consulted our surgical colleague for a patch of cellulitis at the base of his right great toe 5 days ago. Patient is a known diabetic on oral hypoglycemic agents for the last 10 years. He was given linezolid, cefixime, serratiopeptidase and diclofenac by the surgeon. A day after, he developed abdominal pain, hence diclofenac was stopped and symptomatic treatment with pantoprazole, antacid syrup and buscopan was given. Two days later, he developed skin rash on legs. Thought to be due to allergy, hence he was referred for physician consultation.

When I examined him in outpatient department, he was hemodynamically stable. He looked a little pale, abdomen was mildly distended with vague tenderness all over. Petechiae were there all over the lower limbs up to thigh and even on the scrotum (Fig. 1). Scrotum was swollen and tender, ankles were a bit swollen too. Ankle and knee joint movements were painful. He was admitted to the hospital and treated symptomatically with intravenous fluids, pantoprazole, ondansetron, etc.

Investigations

Normal: Hemoglobin, hepatitis B surface antigen (HBsAg), white blood cell (WBC) count, peripheral smear, amylase, prothrombin time (PT) and activated partial thromboplastin time (aPTT), complement 3 and 4 (c3c4), creatinine 1.2 (0.6-1.4).

Negative: HBsAg, antinuclear factor (ANF), hepatitis C virus (HCV), R factor, antineutrophilic cytoplasmic antibodies (ANCA) with cytoplasmic staining pattern cytoplasmic ANCA (cANCA) and perinuclear ANCA (pANCA).

Abnormal: Erythrocyte sedimentation rate (ESR) 30 (0-15), C-reactive protein (CRP) 6.5 (0-6 mg), platelets 4.5 lakhs (1.5-4.0).

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Figure 1. Purpura over feet.

In the view of abdominal pain, joint pain, petechiae over legs HSP was suspected and intravenous hydrocortisone 100 mg thrice a day was started. In spite of the above measures his abdominal pain persisted. Contrast-enhanced computed tomography (CECT) and angiography of abdomen were done. CT showed trasmural thickening of jejunum and terminal ileum suggestive of inflammation/infection (Fig. 2).

He had vomited blood 3 times each 100-150 mL. Upper gastrointestinal (GI) endoscopy showed inactive chronic duodenal ulcer and diffuse gastropathy suggestive of vasculitis (Fig. 3).

He was started on methylprednisolone 1 g every day for 3 days. Patient improved clinically, abdomen scrotal and joint pain resolved. He was discharged on 1 mg/kg prednisolone to be tapered off gradually. He had significant proteinuria of 3 g/24 hours. His blood sugar was controlled with insulin both within the hospital and at home.

He was given telmisartan 40 mg once a day in view of proteinuria. Fundus examination did not reveal retinopathy. Renal biopsy deferred by the patient and skin rash did not reappear during observation.



Figure 2. Edema of intestines on CT scan of abdomen.

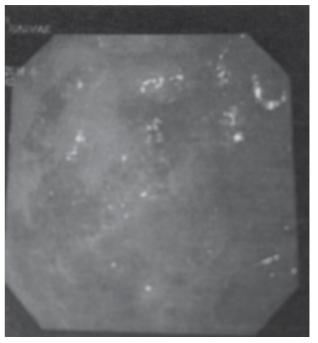


Figure 3. Vasculitis lesions of stomach in upper GI endoscopy.

DISCUSSION

Our patient had focal cellulitis at the base of great toe for which he was given medication. Many drugs are known to precipitate HSP, one of them being diclofenac. He developed skin rash over the leg, which was thought to be simple allergy a usual mistake. He had arthralgia of knees and ankles. HSP often presents with arthralgia before or after onset of skin rash. He had edema of feet, a feature of HSP.

Scrotal involvement in adults is rare.³ Painful scrotal swelling in HSP may require scrotal Doppler study

to exclude torsion of testis where decreased arterial flow points to torsion. Persisting and significant pain abdomen can be due to many a causes in adults with HSP including pancreatitis, appendicitis, cholecystitis, peritonitis or mesenteric vascular occlusion.⁴

Our patient's CT abdomen showed transmural thickening/edema of jejunum and the ileum suggestive of inflammation due to HSP.⁵ CECT scans also help to exclude other causes of pain abdomen. In HSP, intussusception is not a common presentation in adults. Obvious upper GI bleed is rarely seen in adults with HSP, though 56% of children may have occult blood positive stool.

Upper GI endoscopy showed classical diffuse gastropathy suggestive of vasculitis. Sometimes, acute abdomen or upper GI bleed may be preceded by skin rash giving a diagnostic dilemma. GI symptoms without rash also have been reported. Patient responded to high dose methylprednisolone.⁶

In similar situation in nonresponders, immunosuppression, plasmapheresis have also been attempted. Patient had significant proteinuria, as patient had diabetes for a long time and had no diabetic retinopathy; diabetic nephropathy cannot be excluded without renal biopsy, which was deferred by the patient. Proteinuria decreased with prednisolone and telmisartan during observation.

CONCLUSION

In palpable purpuric rash, abdominal and joint pain, with or without renal involvement, we have to consider HSP in differential diagnosis even if the patient is an adult. They may present with upper GI bleed, which needs cautious approach as involvement of GI tract can be diffuse and obscure. Scrotal involvement is rare but a known complication of HSP.

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Traumatic Brain Injuries in Sports

When a purely scientific advance stands to jeopardize a very powerful interest, rejection can turn threatening. Such was the case of forensic pathologist Bennet Omalu, a native Nigerian working in the Allegheny County coroner's office. Dr Omalu had no idea just how powerful the National Football League (NFL) was when he published the first diagnosis of chronic traumatic encephalopathy in Neurosurgery. The NFL immediately mobilized a cadre of physicians on the organization's payroll to attack his research, but Dr Omalu persisted despite such prosecution by the NFL. Even experts without any ties to the NFL initially discounted his research. Because of Dr Omalu's persistence, the NFL has been forced to acknowledge chronic traumatic encephalopathy, and the wider sports culture has begun questioning the costs of repeated brain injuries in sports, both professional and recreational.