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DIAGNOSTIC AND MANAGEMENT CHALLENGES IN A CASE OF HENOCH–SCHONLEIN PURPURA FOLLOWING LAPAROSCOPIC APPENDECTOMY IN A CHILD

General Surgery

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ABSTRACT

Severe Henoch–Schonlein purpura (HSP) is a multisystem, self limiting, non-granulomatous, autoimmune Ig-A mediated small vessel vasculitis mostly affecting children between 4 and 11 years of age. The aetiology is not clearly known, but it is found associated with recent infection, especially respiratory tract, gastroenteritis, food allergies, chemical toxins, vaccination, insect bites, tumors, use of drugs like NSAIDs, antibiotics like penicillin, sulfonamides and ACE inhibitors and alpha-1 antitrypsin deficiency. We report the diagnosis and successful management of severe HSP in a 12 year female child noticed on 2nd postoperative day of laparoscopic appendectomy for acute appendicitis.

KEYWORDS

Acute appendicitis, laparoscopic appendectomy, Henoch-Schonlein purpura, arthralgia, hemetemasis, nephritis.

INTRODUCTION

HSP or Ig-A vasculitis is a systemic vasculitis affecting multiorgan system like skin, joints, GI tract and kidneys. The pathogenesis involves deposition of Ig-A and often other Ig and C-3, in the wall of cutaneous vessels, vessels of renal mesangium and detection of Ig-A immune complexes in the circulation[1]. The clinical presentation includes small erythematous palpable purpura, arthritis, renal and GI menifestations. Pain abdomen is the most frequent GI symptom, while others are nausea, vomiting, hemetemesis, melena and hematochezia. Often the abdominal pain precede the purpuric skin rash typically starting in the lower extremity and the buttocks, which occurred in our case. Severe GI complications due to HSP can be intussusception, ischemic necrosis of bowel, massive GI bleeding presenting as hemetemesis or melena, intestinal perforation, hemorrhagic ascites, pancreatitis, biliary cirrhosis and acute acalculous cholecystitis[2].

CASE STUDY

A 12 years female child brought to surgery OPD with the compliant of pain in the right lower abdomen, nausea, 3 bouts of vomiting and fever since 2 days. On examination, she is thin built, febrile, anicteric, P/A-soft, tenderness in the right iliac fossa with positive rebound tenderness and no palpable lump. Her temp.- 100.2^{0}F, heart rate – 84/min, B.P.- 100/68 mmHg. She had no associated co-morbidity, recent infection or drug intake. She was admitted with the provisional diagnosis of acute appendicitis and started nil by mouth, IV fluids, antibiotics , antiemetics and analgesics. Blood reports revealed a raised TLC of 27,890/cumm with 86% neutroplilia, normal rest of CBC parameters, Hb – 10.9 gm/dl, total platelet count – 3.33 lakh/cumm, RBS, blood urea, serum creatinine and serum electrolytes, serum amylase were within normal limits. Serum lipase was raised 101U/L, CXR was normal. USG abdomen showed a blind ended, non-compressible, aperistaltic tube of 7.5mm diameter, arising from base of cecum with gut signature and inflammed perienteric fat and an appendicolith of size 4.4mm. Free fluid noted in right iliac fossa and pouch of Douglas. Emergency laparoscopic appendectomy done and the intraoperative findings were an inflammed, tortuous, retrocecal appendix surrounded by adhered inflammed and dilated small bowel loops with about 10 ml of thick pus at the tip of the appendix. Rest of the abdominal visceras appeared normal. Her immediate postoperative period was uneventful, but on 2nd post-operative day (POD) she developed erythematous non-itchy papular skin rash over both upper and lower extremities, feet and buttocks and arthralgias of both knees and ankles which became more pronounced on 3rd POD with additional edema of both legs and feet. At this point, the father of the child volunteered about similar skin rash and arthralgias 7 months back, that subsided within a week with some medication from local doctor. There was ongoing mild abdominal pain. Urine (RE and ME) showed – protein ++++(500mg/dl), sugar-nil, pus cell- 10-12/HPF, RBCs- 12-15/HPF, bacteria-nil. On dermatology and pediatrician consultation, suspicion regarding HSP nephritis made and she was transferred to pediatric ward for further management. On POD-4 reports of blood C/S- negative, stool for occult blood- positive, urine C/S- grew Enterococcus species sensitive to Linezolid and Vancomycin. On POD-5, serum p-ANCA and c-ANCA for Crohn's disease were negative. On POD-6, complement protein concentration (C3 and C4) were normal, spot urine protein was very high at 106.9 mg/dl (normal <15mg/dl), spot urine creatinine was very low at 4.7mg/dl (normal 28-217mg/dl) and a very high protein creatinine ratio of 22.74 mg/dl (normal <0.2). From 4th POD, she was started Inj Hydrocortisone 100mg IV bolus followed by Inj Dexamethasone 8mg IV OD for 5 days and then 4mg IV OD for 2 days. On 6th POD she had 2 episodes of hemetemesis and one bout of hematochezia. She was transfused one unit of packed RBCs and managed in paediatric ICU for 2 days. However she recovered fully after 5 days of injectible steroid, injectible antibiotics and supportive therapy. She tolerated normal diet and got discharged on 12th POD after removal of port site sutures, On POD-11, she was given Tab Prednisolone 20mg PO, OD which was tapered gradually over 2 weeks and stopped. The papular skin rash resolved completely by 20th POD. The child was followed monthly and doing well till 8 months and without further relapse.

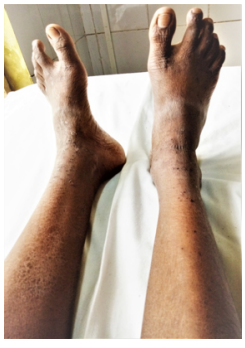

Figure 2 : Nonpalpable purpuric lesions over both leg and feet.

DISCUSSIONS

HSP is commonly encountered in children below 10 years of age in up to 90% cases with an overall incidence of 2 per 1000 children[3]. The mainstay of diagnosis of HSP is clinical, as laboratory tests are non-specific unlike other types of vasculitis. The clinical menifestations are variable with usual early occurrence of palpable purpuric skin rash, commonly over the lower limbs and buttocks than upper extremity and other sites[4]. This Ig-A mediated small vessel vasculitis predominantly involves skin, intestine and renal glomeruli and manifests clinically as palpable purpura, abdominal colic, hematuria and arthralgia[5]. An occasional patient may develop facial and periorbital edema, scrotal edema and bilateral pedal edema. Vasculitis of the ileocecal region can mimic clinically as appendicitis with resultant un-necessary abdominal exploration and appendectomy reported in about 5-7% cases[2]. Patients with abdominal pain or other GI menifestations at the start can occur in 12-19% cases, few days prior to the onset of skin rashes posing a real diagnostic difficulty[6]. USG and CECT of the abdomen, in equivocal cases of right lower abdominal pain settle the issue of terminal ileitis from appendicitis and helps preventing un-necessary surgery. However, a postoperative case of exploration of abdomen or appendectomy when suffers a stormy course of continuation of abdominal pain, development of skin rash in lower limbs and buttocks, arthralgias, hematuria or protenuria should raise a strong suspicion to the underlying HSP as in our case. Atypical and rare HSP presentations include pulmonary as alveolar hemorrhage and hypoxemia, cardiac and neurological complications[7]. The differential diagnosis of HSP are Crohn's disease, Ig-A nephropathy, Wegener's granulomatosis, hemolytic uraemic syndrome and other vasculitidis syndromes and infections.

A milder form of HSP in children and adults is the common occurrence and usually resolve spontaneously without any specific treatment recommended for it. NSAIDs are frequently prescribed for joint pain after excluding renal involvement and antispasmodics for abdominal colic. All acute and severe form of HSP especially with joint and GI involvements need to be diagnosed by high level of suspicion and exclusion of other differential diagnoses and are treated with IV steroids, i.e. hydrocortisone (100-200 mg OD) or dexamethasone(4-8mg OD) for 3-5 days and then followed by oral prednisolone for few weeks with gradual tapering dosage. Patients with renal involvement needs to be added with immunosuppressive drugs like Azathioprine 2-3 mg/kg/day or Cyclophosphamide 3mg/kg/day or triptolide 20mg thrice daily or mycofenolate mofetil[8] . This aggressive and combined therapy is very helpful in averting end stage renal disease or morbidity and mortality due to GI bleeding. The protenuric patient may be started on angiotensin converting enzyme (ACEs) like Enalapril or Ramipril or Angiotensin receptor blockers (ARBs) like losartan or irbesartan as renal protectors. Hemodialysis may occasionally be needed for a severely deranged renal function especially in an adult.

HSP is self limiting in up to 94% cases in children and 89% cases in adults[2]. Overall prognosis of HSP is good and usually improvements occur within 6-8 weeks and is associated with a 5 year survival rate of 95%[9] . Relapses due to HSP occur in 30% cases usually within 4 months with lesser severity and shorter duration of illness[9]. A poorer prognosis occurs with HSP onset at delayed age, severe skin involvement above waist line, immunoglobulin imbalance and neurological involvement.

CONCLUSIONS

HSP is an uncommon, non granulomatous Ig-A induced vasculitis, mostly self limiting and manifesting as non thrombocytopenic palpable purpura with multiorgan involvement including joints causing arthralgias, GI tract, kidney causing glomerulonephritis, hematuria and protenuria. The diagnosis of HSP is difficult and needs a high clinical suspicion and judgement as laboratory tests are only supportive. It is common in younger children with a better prognosis, while late age onset and delayed presentation with severe disease leads to higher morbidity and mortality. Abdominal pain is the commonest GI symptom and can mimic appendicitis and may result in an unnecessary abdominal exploration and appendectomy in most of the cases. In few cases, within 2 to 3 days of surgery there appears the classical menifestations of palpable skin rash, pedal edema, arthralgias, GI symptoms, hematuria or protenuria. However only few cases have been reported to have appendicitis due to HSP requiring appendectomy and HSP diagnosed in the early postoperative period as in our case. Hence a high level of suspicion in such difficult case scenarios can help early diagnosis, emergent management and thus preventing life threatening complications of HSP.

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