

Perioperative management of a patient with Henoch–Schonlein purpura for appendectomy

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ABSTRACT

Henoch-Schonlein purpura (HSP) is a multisystem disease and immunoglobulin A-mediated vasculitis with a self-limited course affecting the skin, joints, gastrointestinal tract, and kidneys. It is the most common form of acute small-vessel vasculitis primarily affecting children. Severe renal and central nervous system disease may lead to life-threatening conditions, and immunosuppressive agents and plasmapheresis may be needed. The cause of HSP is unknown; immunizations, certain food allergies, insect bites, infection, and some medications may play a role in the development of the disease. Perioperative management for liver and kidney functions is very important for anesthetized patients with HSP. We report the perioperative management of a patient with HSP for appendectomy.

Saudi Med J 2006; Vol. 27 (5): 714-716

Henoch-Schonlein purpura (HSP) is the most common form of acute small-vessel vasculitis primarily affecting children. Clinical features include skin rashes, arthritis, abdominal pain, and nephritis. Gastrointestinal (GI) symptoms occur in up to 85% of the patients. It occurs most often in children between the ages of 3-10 years, and presents classically with a unique distribution of the rash to the lower extremities and the buttock area.¹⁻² Severe renal and central nervous system disease may lead to life-threatening conditions, and immunosuppressive agents and plasmapheresis may be needed.³ Renal involvement is manifested by hematuria and proteinuria. Treatment is symptomatic. The overall prognosis in HSP is excellent, but the long-term morbidity depends on the renal and neurologic involvement. One third to one half of patients will have one or more recurrences of symptoms, usually within 6 weeks, but may occur as late as 3-7 years later.⁴ The cause of HSP is unknown, immunizations (smallpox vaccination), certain food allergies (milk, egg, chocolate, wheat), insect bites,

infection (influenza, β -hemolytic *Streptococcus*), and some medications (penicillin, quinine, chlorothiazide) may play a role in the development of the disease. Perioperative management for liver and kidney functions is very important for anesthetized patients with HSP.

Case Report. A 12-year-old male, weighing 42 kg, diagnosed with HSP was admitted to our hospital. He has no history of allergy and systemic disease. Earlier, he developed arthralgias and palpable purpura that were distributed over the buttocks and the posterior aspect of arms and legs. A diagnosis of HSP was confirmed, and he was started on methylprednisolone 45 mg a day. He presented with colicky abdominal pain for one day. The pain was periumbilical in location, associated with recurrent bilious vomiting and obstipation. On examination, he was pale and in agony; he had diffused purpura over his buttocks and lower extremities (**Figure 1**), pulse rate was 98/minute,

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Received 18th September 2005. Accepted for publication in final form 31st December 2005.

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blood pressure 105/70 mm Hg, peripheral oxygen saturation 98%. Abdominal examination revealed diffuse tenderness, mild guarding but no rigidity. There were no organomegaly, and bowel sounds were exaggerated.

Laboratory examination revealed white blood cell (WBC) 7500/uL; hemoglobin (Hb) 11.4 g/dL; hematocrit (Hct) 35.4%; platelets (Plt) 321.000 uL; neutrophils (Ne) % 75.6% (normal ranges: 40-74%); lymphocytes (Ly) % 14.8% (normal ranges: 16-45%); erythrocyte sedimentation rate 68 mm/1.h. Liver and kidney function tests, serum electrolytes, calcium and magnesium examination did not show any abnormality. Urea: 28.5 mg/dl (normal values 10-50 mg/dl), blood urea nitrogen: 13.36 (normal values 7-20), Cre: 0.6 mg/dl (normal value 0.6-1.3 mg/dl), aspartate aminotransferase: 19.04 U/L (normal values 0-32 U/L), alanine aminotransferase: 14.56 U/L (normal values 0-41 U/L), Na: 137.9 mmol/L (normal value 136-145 mmol/L), potassium: 4.36 mmol/L (normal values 3.5-5 mmol/L). Prothrombin time and partial thromboplastin time levels were normal. Urine examination showed 7-10 RBC/hpf, urine protein: 24 mg/d (normal values \leq 150 mg/d) (5.2/mg/m²/24 h.). The stool for occult blood was negative. Serum immunoglobulin (Ig) levels were normal: IgG: 12.3 g/l (normal values 5.50-19.00), IgM: 0.612 g/l (normal values 0.55-1.45), IgA: 2.37 g/l (normal values 0.60-3.30), IgE: 12.32 unit/ml (normal values 12-681), anti-streptolysin: 174 u7ml (normal values 0-200), C-reactive protein : 29.3 mg/l (normal values 0-8), complement 3 (C3): 1.3 g/l (normal values 0.790-1.52), C4: 0.263 g/l (normal values 0.160-0.300). Chest x-ray was normal. X-ray of the abdomen revealed air-fluid levels. The diagnosis of the ultrasonography of abdomen was acute appendicitis.

After premedication with intravenous (IV) 2 mg midazolam, he was admitted to the operating room. Propofol 120 mg, fentanyl 50 μ g, and rocuronium 20 mg were used in the induction. Tracheal intubation was performed. Anesthesia was maintained with sevoflurane 2-3%, in 50% O₂ + 50% N₂O. Intraoperatively, and in the following 2 days and remained in the intensive care unit, no problems occurred. Repeat blood count on the first postoperative revealed: WBC 6600/Ul, Hb 10.1 g/dL, Hct 31.2%, Plt 255.000 uL, Ne% 75.6%, and Ly% 14.8%. Repeat urine examination showed: urine protein 4.3 mg/m²/h. He improved clinically and investigation reports normalized.

Discussion. Henoch-Schonlein purpura, also known as common childhood vasculitis, was first described in 1837. The symptoms include skin



Figure 1 - Henoch-Schonlein purpura on the lower extremities.

rashes and arthritis and Schonlein named it as peliosis rheumatica. In 1868 Henoch described GI as one of the symptoms of HSP.⁵ Henoch-Schonlein purpura is a multisystem IgA mediated vasculitis with a self-limited course affecting the skin, joints, gastrointestinal tract, and kidneys. Skin biopsy on immunofluorescence often reveals granular depositions of IgA and C3 within the walls of the dermal vessels as well as in the connective tissue of the upper dermis.⁶ The American College of Rheumatology published diagnostic criteria for HSP in 1990,⁷ including age less than or equal to 20 years at disease onset, the presence of palpable purpura, GI bleeding, and a biopsy showing granulocytes in the walls of small arterioles or venules. The diagnosis of HSP can be difficult, especially when abdominal symptoms precede cutaneous lesions. A skin rash is nearly always present, although not necessarily in the earliest stages. Normal platelets count, prothrombin time, and partial thromboplastin time excludes a platelet or hemorrhagic disorder as the cause of purpura. Other laboratory tests are usually not conclusive.

Acute pediatric abdomen is a very common clinical problem. Abdominal symptoms reportedly occur in 50-85% of HSP patients, possibly secondary to edema and hemorrhage within the bowel wall and mesentery. The abdominal pain is always colicky and poorly localized. On physical examination, the abdomen may be rigid or distended, occasionally mimicking an acute abdomen and resulting in unnecessary exploratory laparotomy. Signs suggestive of intussusception including an abdominal mass may be present in children. The GI bleeding occurs in one-third of cases and may be overt, with hematemesis, melena, or bloody stools. While bloody diarrhea is common in HSP, it may also occur in eosinophilic gastroenteritis, systemic lupus erythematosus, parasitic

infection, and drug-induced vasculitis. Distinguishing between these different vasculitic disorders depends on clinical, serological, hematologic, and histologic findings. Arthralgia is usually symmetrical, with the ankles being the most commonly affected joints. Joint involvement is considered as an indication for systemic steroids. Proteinuria and hematuria indicate renal involvement and generally occur within the first 3 months after the onset of the disease.⁸

The diagnosis of HSP may be difficult, especially when abdominal symptoms precede the characteristic palpable purpura. Clinical and laboratory findings, however, are nonspecific or confusing in many instances. Selection of an appropriate imaging modality is essential to ensure prompt management. In the majority of cases, ultrasound can provide specific diagnoses, whereas in others valuable supplemental information can be obtained. Computed tomography (CT) will be reserved for selected patients in whom further information is needed. Indications for magnetic resonance imaging (MRI) in the management of acute pediatric abdomen are currently limited. The MRI, however, is indicated on an emergency or semi-emergency basis in selected conditions including anomaly of the internal genitalia, ovarian torsion, and congenital biliary dilatation.⁹ The hallmarks of HSP on CT are multiple focal areas of bowel thickening with skip lesions, mesenteric edema, vascular engorgement, and non-specific lymphadenopathy. However, similar thickening may also be seen in eosinophilic gastroenteritis, lymphoma, Crohn's disease, and granulomatous disease. It is thus, not a specific sign for HSP. Ultrasound may reveal generalized thickening of the intestinal wall, ascites, and sometimes intussusception.

The GI involvement in HSP is seen predominantly in the small bowel but may also affect the esophagus, stomach, terminal ileum, and colon. Our patient had diffused, poorly localized abdominal pain and bloody diarrhea, prompting us to visualize both the upper and lower GI tract. The endoscopic findings include discrete coin-like petechiae, hemorrhagic erosions, and skip hyperemic and ecchymotic lesions. These were seen in the gastric antrum, cecum, ileocecal valve, and sigmoid colon. It is important to recognize these characteristic endoscopic findings, when a previously healthy patient present with the sudden onset of an acute abdomen. When encountered, they should suggest the possibility of HSP. Although, we did not biopsy the lesions in the GI tract, we expected to find histologic abnormalities similar to those found on skin biopsy. However, endoscopic biopsies often miss the submucosal vessels and only reveal nonspecific inflammation.¹⁰ The HSP is primarily a medical

disease and requires only supportive treatment once other acute surgical conditions have been excluded.¹¹ Simple analgesics or non-steroidal anti-inflammatory drugs are used as first-line therapy for relief of arthralgia. Parenteral steroids have been advocated for more severe abdominal or joint pains, or painful angioedema, but they should be used with caution if there is active GI bleeding. All patients with HSP should have their urine analyzed on several occasions during the initial stages of the disease. Proteinuria and hematuria indicate possible renal involvement, which if progresses to renal insufficiency, has a poor long-term outcome. Our patient's urinalysis was hematuria.

In conclusion, although the cause of HSP is unknown, perioperative management for liver and kidney functions is important for anesthetized patients with HSP. Sufficient IV fluid administration is necessary. Attention should be paid to decrease the risk of tissue compression such as that associated with positioning, blood pressure cuff, and endotracheal intubation, which may cause necrosis over pressure points.

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