CASE REPORT

Henoch - Schonlein Purpura

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ABSTRACT. Henoch-Schonlein purpura (HSP) is an immunologically mediated systemic vasculitis of small blood vessels that primarily, involves skin 100%, gastrointestinal tract 50-70%, joint 70% and the kidney 20-100%. The most common clinical manifestation are intermittent purpura, athralgia, abdominal pain and hematuria. The diagnosis of HSP based on clinical manifestation, laboratory finding and skin biopsy. The manifestation of HSP in a case of eleven year old Balinese girl were maculo papular rash of the flexor, and extensor region of the legs, butocks and fore arms, abdomininal pain and bloody stools Laboratory finding were WBC 20,000/µl, platelet count 490,000/µl, Bleeding time 3 minutes, clothing time 14 minutes PTT 23,7 second, Prothrombin time 11,2 second. The blood urea nitrogen, 16 mg/dl, creatinine, 0,66 mg/dl. Complemen C4, 39 mg/dl, IgA 355 mg/dl ASTO and CRP were negative. Histologic examinitation showed epidermis with hiperkeratosis, proliferation of subepidermic conective tissue, with magnitude of leucosites infiltration surounding small blood vesels with the conclution was according to feature of HSP. **[Paediatr indones 1997;37: 86-90]**

Introduction

Henoch-Scholein purpura (HSP) is an immunologically mediated systemic vasculitis of small blood vessels that primarily, involves skin 100%, gastrointestinal tract (50-70%), joints (70%), and the kidney (20%-100%).[1-3] The most common clinical manifestation are intermittent purpura, Athralgia, abdominal pain and hematuria.[4-6] Although the incidence of this disorder have not been reported, but HSP is the most common form of hypersensitivity vasculitis in children between the ages of 5 and 15 years.[7,8] It is

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rarely in children under 2 years old and adult, males are affected more often than females. The purpose of this paper is to report a case of Henoch-Schonlein purpura in an eleven year old Balinese girl.

Report of the Case

An eleven year old Balinese girl was admitted to the Department of Child Health, Sanglah General Hospital, on July 29, 1996, with a main complaint of purpuric lesions. One week prior to admission complained of sore throat without fever for two days. A maculopapular rash appeared at the lower limbs and both feet accompanied by the passage of bloody stools for one day. Then she was referred to Negara District Hospital. and was treated with ampicillin and prednison for two days. Because the rash migrate to the upper limbs, buttock and both hands she was referred to Sanglah General Hospital on July 29, 1996. She had had a history of similar skin rash on July 11, 1996, but there was no evidence of bloody stools, hematuria, swelling or pain in any joint, nor food and drug allergy.

On physical examination, she was alert with pulse rate 92 beats/minute, respiration rate, 28/minute body temperature was 36.5° C and the systemic blood pressure was 120/80 mmHg. She was well nourished with body weight of 27 kg. There was no evidence of conjunctival bleeding, anemia or palpebral edema, but she had a mild throat inflammation. The chest was symmetrical, and showed no retraction. Heart and lungs were normal. She had slight epigastric tenderness on deep palpation. Normal bowel sounds were heard in all quadrants. Spleen and liver were not palpable.

There were symmetrical petechiae and purpural rashes, measuring from few millimeters to 1.5 centimeters in diameter with small necrosis in the center of same rash, Above the purpura rash we still see the macula papular rash. Under the purpura rash a brownish desquamation skin lesion replaced the purpura. The skin rash appeared in the flexor and extensor region of the legs, buttocks and fore arms. There was no joint swelling or pain in any of joints.

Laboratory investigation disclosed hemoglobin concentration of 13.4 g/dl; hematocrit, 38%; WBC, 20,600/µl, platelet count, 490,000/µl, bleeding time 3 minutes, clotting time 14 minutes; PTT 23.7 seconds and prothrombin time, 11.2 seconds. The blood urea nitrogen was 16 mg/dl; creatinine, 0.66 mg/dl; total protein, 1,06 g/dl, Albumin, 4,08 g/dl and cholesterol 311 mg/dl. The complement C3 104 mg/dl (N:55-120 mg/dl), C4, 39 mg/dl (N:20-50 mg/dl), and IgA, 355 mg/dl (N:85-450 mg/dl).The erythrocyte sedimentation rate was 4/24 mm in 15 and 45 minutes. On blood smear showed normocytic normochromic anemia. Her urinalysis showed a specific gravity of 1.015 without blood or protein, and the sediment count, 2 to 3 WBC without RBC or cast. Her stool showed evidence of blood and mucus.

The working diagnosis was suspected Henoch-Schonlein purpura. The patient was

then treated with prednison 10 mg every 8 hours. She was given also multivitamin and ampicillin 500 mg every 8 hours. On August 2, 1996, she complained of abdominal pain followed by vomiting and bloody stool. Two days later new purpuric lesions appeared at the initial sites, predominantly on the thigh and buttock. The erythematous papular rash which were appeared previously, became dark brown in color, with desquamation skin lesions. On August 7, 1996, laboratory investigation showed negative anti streptolysin O (ASTO) and C-reactive protein (CRP) but anti nuclear antibody (ANA) test was positive. The throat culture was positive for streptococcus and staphylococcus. The stool and urine were normal, where RBC did not found in both sample.

The patient was consulted to the Department of Dermatology, and was diagnosed of suspected Henoch-Schonlein purpura. Skin biopsy was done on August 14, 1996 and the result showed epidermis with hyperkeratosis, proliferation of subepidermic connective tissue, with magnitude of leukocytes infiltration surrounding small blood vessels. The conclusion was Henoch- Schonlein purpura. The patient was discharged on August 20, 1996. Three weeks after admission, there were no new purpuric lesion developed and the skin lesion has almost completely healed. No RBC was found on urinalysis and stool examination.

Discussion

Henoch-Schonlein purpura (HSP) is a multisystem disorder affecting predominantly the skin, joints, gastrointestinal tract, and kidney, although other organs can rarely involved. Johann Schonlein first described the condition that he called "reliosis rheumatica", in which arthralgia was associated with purpura. Then Edward He- noch described a syndrome of purpura, severe abdominal colic and melena.[2] HSP is also know as non-thrombocytopenic purpura, anaphylactoid purpura, allergic purpura or Henoch-Schonlein syndrome.[2,5,6,8,9]

HSP is thought as a result of an autoimmune disease that produces inflammation or vasculitis in small blood vessels resulting in perivascular infiltration and serosanguineous effusion into surrounding tissues, producing the characteristic purpura.[10] Involved vessels show endothelial swelling often associated with occlusion of the lumen, diapedesis of erythrocytes and fibrin deposition in and around the vessel wall. Fibrinoid degeneration is also common and refers to the changes visible in the vessel wall secondary to edema and fibrin deposition. Cellular infiltrate consisting of PMNL, mononuclear cell, and occasional eosinophil. In patient with normal serum complement levels, lymphocytes were the predominant cells. Immunoflourecence demonstrates complement and immunoglobulin in peripheral and glomerular vessel wall especially in lesions less than 24 hours old.[11] The diagnosis of HSP in this case based on clinical manifestations, laboratory findings and skin biopsy. In this case the disorder involved only two system, i.e., the skin and gastrointestinal tract.

Gastrointestinal manifestation of HSP occur in 50 to 70% of all affected children but the incidence rises to more than 90% in those who have renal involvement. The commonest symptom is abdominal colic, nausea, vomiting, diarrhea, while melena occur in half of the cases.[2,6] In our case that symptoms also found such as, vomiting, abdominal pain and bloody stools for one day. The symptoms of gastrointestinal tract usually cause by extravasation of blood or serosanguinous fluid into the wall of the small intestine, which have been pointed on operation. Edematous, scarlet segmental lesion also have been described.[12]

The cutaneous purpura in this case is similar in predilection to HSP. Papular purpura in HSP appears typically on extension surface of the lower extremities particularly on buttock, shins, and ankle, but it may also present on other part of the body.[1,6,10,13] Chest, neck and face were involved less frequently.[5,13] It usually starts as a symmetrical erythematous macular rash that soon changes to a maculopapular and purpuric form. The purpura may coalesce into large ecchymoses.[1,3,13]

Skin lesion in HSP is unlike to systemic lupus erythematosus (SLE) which is also can involve multisystem. The skin lesion in SLE is predominantly seen at the sun shine exposed skin. The unexposed skin is normal. The most common lesion is butterfly-like appearance specifically in the cheek, can be as a simple erythematous lesion or maculopapular eruption with reddish fine squamous.[11] Other vascular purpura should also be considered. The purpura associated with inherited connective tissue disorders, such as Ehlers-Danlos syndrome, often is accompanied by large ecchymoses and hematomas.[1] The purpura also can be simple but more often is accompanied by swelling and blebs, or diffuse erythematous with or without swelling.[12] The purpura seen with scurvy typically occurs around near follicle and the distribution as a saddle area in the thighs and buttocks. Ecchymoses and large subcutan hematomas may also occur.[10,12] Skin biopsy may helpful in an occasionally case, but essentially non specific finding in HSP.[12] In our case histologic pattern of the skin biopsy was according to HSP.

Laboratory findings also confirm the diagnosis of HSP, where the titer of serum complement C3 and C4; and serum IgA are in the normal range.[1] It is different than SLE in the active stage, where the titer of serum complement C3 and C4 are usually decreased. One of the test which did not support the diagnosis of HSP in our case was the positive result of antinuclear anti body. This test usually positive in SLE.[6] Can it be also positive in HSP? Are there any other mechanisms for tissue injury in HSP, such as the involving of antitissue antibody? In one of the syndrome of hypersensitiviity vasculitis such as urticarial vasculitis, may have a positive anti nuclear antibody (ANA) test, but have low serum complement (C3).[7]

The etiology and the nature of the immunological reaction of HSP is not completely clear. The illness is preceded by upper respiratory infection in 30 to 50% of patients. HSP has been reported to follow streptococcal infection, smallpox vaccination, insect bites, exposure to cold, drug ingestion and food allergy.[2,9] In this case the agents which

have closed association to the HSP were streptococcus and staphylococcus, isolated from throat swab culture. Besides vascular factor, purpura can also be caused by platelet and coagulation defects. But this possibility can be excluded by the normal results of platelets count, bleeding time, clotting time, PTT and prothrombine time.[5]

The overall prognosis of HSP is better appreciated from two large series in which both with and without nephritis were follow up giving estimate incidences of end stage renal failure of 2 % and 5%.[2]

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