

Two Cases of Henoch-Schönlein Purpura with Transient Myocardial Ischaemia

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Sir,

Henoch-Schönlein purpura is a systemic vasculitis that affects small blood vessels. The clinical symptoms consist of palpable purpura, mainly on the extensor surfaces of the lower limbs, in association with joint, gastrointestinal or renal involvement to varying degrees. There have been only a few reports of Henoch-Schönlein purpura with cardiac involvement (1–4). We describe two young men with Henoch-Schönlein purpura who developed transient myocardial ischaemia in the course of the illness. It is likely that epicardial coronary spasm, which occurred in association with systemic vasculitis, gave rise to the events in our patients.

CASE REPORTS

Patient 1

A 28-year-old man was admitted to our hospital because of recurrent, purpuric eruption on the lower extremities with a history of 3 months. In addition, he had been suffering from mild intermittent pain on the upper abdomen for 3 days prior to admission to hospital. His medical history was unremarkable and he did not smoke cigarettes.

On examination, a large number of palpable, purpuric lesions were found scattered on the dorsa of the feet, extensor aspects of the legs, and buttocks (Fig. 1). He was not obese, and his blood pressure was normal. Normal or negative laboratory investigation included urinalysis, blood cell count, chemistry panel, including plasma electrolyte, plasma lipoproteins, fasting blood sugar and serum immunoglobulin and complement levels. Anticardiolipin antibodies were not detected. Stool was positive for occult blood. Bacterial cultures of the throat yielded streptococcus pyogenes. A biopsy specimen from a purpura on the lower leg revealed a leucocytoclastic vasculitis with IgA and C3 deposition along the vascular wall in the upper dermis.

An electrocardiogram taken on admission showed ST elevations and inverted T-wave in leads V_2 – V_5 , suggesting anterior-wall ischaemia. Hyperventilation was excluded for the normal results of blood gas analysis. Serum creatine phosphokinase was not elevated. He denied having chest pain or discomfort despite repeated questioning. Echocardiography showed no abnormal change. Pericardial fluid was not detected. Administration of Ca-antagonists was started and the electrocardiogram returned to normal 5 days later. The treadmill test resulted negative. Coronary angiography was not performed.

The abdominal pain resolved spontaneously within several days without any particular medication. The purpura decreased gradually, finally disappearing 8 months later, but a mild proteinuria (below 1 g/day) developed in the course of the disease. He has not complained of chest pain or discomfort, and the electrocardiograms taken regularly in the following 2 years have not shown any abnormal trace.

Patient 2

A 20-year-old man was transferred to our hospital from an outside hospital because of intermittent, severe, colicky pain on the upper abdomen continuing for 10 days. His past medical history was unremarkable. He did not smoke cigarettes. Several days after admission, a large number of palpable purpura occurred on the lower extremities, spreading over the buttocks and arms. He was not obese, and his blood pressure was normal. Histology of a biopsy specimen revealed a leucocytoclastic vasculitis with IgA deposition along the vascular wall in the upper dermis. Laboratory studies revealed the following abnormal values: leucocyte count 10,700 cells/mm³, with 78.2% neutrophils, C-reactive protein 5.4 mg/dl (normal <0.4), and occult blood test of the stool 2000 ng/ml (normal <100). Urinalysis, chemistry panel, including plasma lipoproteins, fasting blood sugar and serum immunoglobulin levels were within normal limits. Anticardiolipin antibodies were not detected. Upper gastrointestinal endoscopy revealed multiple ulcers in the descending duodenum. An electrocardiogram gave a normal tracing. Prednisolone 60 mg daily was started, resulting in a remarkable improvement of abdominal symptoms within 2 days. Since the purpura subsided gradually, the dose of prednisolone was tapered to 40 mg over 5 weeks. Two days after the dose of prednisolone had been tapered to 40 mg, he complained of retrosternal pain in the morning while resting in bed. The pain lasted approximately 30 min. He described the nature



Fig. 1. Palpable, purpuric lesions on the lower legs in Patient 1.

of chest pain as heaviness. The electrocardiogram taken at that time revealed an inverted T-wave in lead III, and a flat T-wave in leads II and a_{VF} , suggesting inferior-wall ischaemia. Serum creatine phosphokinase and plasma electrolyte were within normal limits. An echocardiogram was interpreted as normal. The abnormal change in the electrocardiogram returned to normal within 4 days. Coronary angiography was not performed. The patient has not had any coronary events for 2 years, although no coronary vasodilators have been administered. He is now doing well, taking a daily dose of 2 mg of prednisolone without any symptoms of Henoch-Schönlein purpura.

DISCUSSION

Only a small number of patients with Henoch-Schönlein purpura associated with cardiac involvement have been reported in the literature. Lecutier (1) described an 11-year-old boy with Henoch-Schönlein purpura who died approximately 1 month after onset of the disease. A throat culture yielded streptococcus pyogenes. Post-mortem examination revealed several areas of necrosis with calcification in the myocardium, but neither coronary arteries nor arterioles perfusing the myocardium were involved. They suggested that the antigen-antibody reaction might affect the myocardium in the same way as it produces the vascular lesions. Subsequently, Imai & Matsumoto (2) reported a 14-year-old girl with Henoch-Schönlein purpura, developing heart failure about 2 weeks after the onset of her illness. The electrocardiogram indicated myocardial injury compatible with an acute carditis. The antistreptococcal titre was markedly raised.

Coronary events in the course of the disease have also been described (3, 4). It is likely that our two patients developed transient myocardial ischaemia in the course of Henoch-Schönlein purpura. Acute myocardial infarction was excluded in both cases, because ischaemic changes in the electrocardiogram returned to normal within 4 to 5 days, and the level of creatine phosphokinase was not elevated. Acute pericarditis was also excluded in Patient 1, because elevation of ST segments was confined to precordial limbs in the electrocardiogram, and the echocardiography revealed no pericardial effusion. Neither patient had atherosclerotic risk factors such as obesity, cigarette smoking, hypertension, diabetes mellitus or abnormalities in plasma lipoproteins. Thus, it is not likely that there were significant epicardial coronary stenoses due to atherosclerosis in our patients, although the absence of this finding could not be documented by coronary arteriography. Coronary events occurred at rest in both patients, and the result of the treadmill test was negative in Patient 1. On the basis of these facts, it seems likely that coronary spasm caused transient myocardial ischaemia in our patients.

In Patient 1, the episode of myocardial ischaemia was not associated with chest pain or discomfort. We detected it coincidentally from the abnormal tracing on the electrocardiogram taken on admission. Continuous electrocardiogram recording reveals that about 80% of episodes of transient myocardial ischaemia do not cause chest pain or any other symptoms (5). This might explain the absence of reports on transient myocardial ischaemia in association with Henoch-Schönlein purpura.

The mechanism for the development of coronary spasm in our two patients remains to be elucidated. One possible explanation is that systemic angitis induced endothelial damage which sequentially released vasoconstrictive substances from those cells, such as endothelin-1 known to have potent vasoconstrictive actions, leading to the spasm of coronary arteries. The plasma endothelin-1 levels have been shown to elevate in a variety of systemic vascular diseases (6). Toyo-oka et al. (7) demonstrated a significant change of plasma endothelin-1 levels in coronary sinus blood during and after coronary spasm provoked by intracoronary administration of acetylcholine or ergonovine, suggesting that this potent vasoconstrictor agent may play an important role in coronary spasm.

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