Myocardial infarction – a rare complication in Henoch-Schönlein purpura

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Summary

A 29-year-old man with previous Henoch-Schönlein disease presented with multiple systemic emboli and a myocardial infarction. Subsequent investigation by angiography showed normal coronary arteries. This appears to be the first reported case of Henoch-Schönlein disease and myocardial infarction probably due to coronary vasculitis.

Introduction

Henoch-Schönlein disease (anaphylactoid purpura) is a clearly defined syndrome. It is characterized by non-thrombocytopenic purpura, polyarthritis, localized subcutaneous oedema, glomerulonephritis and gastrointestinal manifestations including bleeding and small bowel obstruction. Its aetiology is unknown but circumstantial evidence implicates hypersensitivity to bacteria, viruses and food (Robinson, 1977). All ages may be affected but the syndrome is more common in children (median age 4 years). The underlying pathological process is vasculitis with perivascular accumulation of polymorphs and erythrocytes, together with glomerular deposition of immunoglobulins, particularly IgA and complement. The clinical picture is thought to reflect the severity and site of immune complex deposition (Cream, Gumpel and Peachey, 1970).

Case report

A 27-year-old Caucasian bricklayer presented in 1977 with polyarthritis, haematuria and purpura on the extensor surfaces of all limbs. Investigation and clinical course led to the diagnosis of Henoch-Schönlein purpura.

Right renal biopsy performed 6 months after his original admission showed a severe focal and segmental proliferative glomerulonephritis; 55% of the glomeruli were obsolete and most of the remainder showed a segmental increase in the number

of mesangial cells and increase of mesangial matrix. Coarse granular deposits of IgA, IgM and C₃ were present in the mesangium. On electron microscopy dense subepithelial and subendothelial deposits and smaller intramembranous and mesangial deposits were present. There was focal involvement of arterioles by fibrinoid necrosis. The appearances were consistent with Heboch-Schönlein nephritis.

In 1979 he had an episode of severe precordial pain lasting 24 hr and unrelieved by rest but which resolved spontaneously. Two months later he had a similar, milder episode. On neither occasion did he seek medical attention. One month later he developed colicky abdominal pain and numbness in the left leg. Blood pressure at this time was 120/70 mmHg. ECG showed a previous inferior myocardial infarction. Laparotomy performed because of radiological evidence of small bowel obstruction was normal. Aortography was performed. Left femoral embolectomy removed recent thrombus from the bifurcation of the superficial and profunda femoris arteries. The clot was histologically normal and sterile on culture. Postoperatively, the patient developed acute renal failure which subsequent urography and renographic scanning showed was due to a left renal embolus. While recovering he developed the sequential ECG and enzymatic changes of an antero-lateral myocardial infarction.

Investigation showed a normal full blood count, electrolytes, liver enzymes, glucose tolerance and cholesterol. Triglycerides were 2.66 mmol/l and immunoglobulins were normal apart from IgG of 5.1 gm/l. The ESR was 50 mm/hr. Rheumatoid factor and antinuclear factor were absent and DNA binding less than 20%. Complement profile CH₅₀: 135% normal, C₃ 103% normal, C₁ binding immune complexes present in the serum which contained IgM. Blood urea was 8 mmol/l, urinary protein 3 gm/l and 24-hr creatinine clearance was 37 ml/min.

Exercise testing, echocardiography and ambulatory monitoring were unremarkable. Four months after his first admission ventriculography showed mild apical dyskinesia but coronary angiography was entirely normal (Fig. 1).

Discussion

Acute myocardial infarction with normal or near

normal coronary arteries is a well established entity (Dear *et al.*, 1971; Potts, Stein and House, 1972). Several factors have been implicated – pulmonary embolism, calcific aortic valve stenosis, severe acute anaemia, hypotension and malignant hypertension – but in the majority of cases the aetiology remains unknown. Characteristically, there are few risk factors, no preceding angina and, on coronary angiography, the vessels are found to be normal.

FIG. 1. Coronary angiograms demonstrating normal appearances of left main stem, anterior descending and circumflex arteries (a) and right coronary artery (b).

Several mechanisms have been proposed: recanalization or resolution of an *in situ* thrombus, coronary artery emboli, coronary artery spasm, abnormal intramyocardial vessels, abnormal haemoglobins affecting oxygen transport and platelet aggregates with prolonged disaggregation time. A combination of coronary artery spasm (Cheng *et al.*, 1972) and platelet aggregation (Oliva and Brechinnidge, 1977) has been postulated as fundamental in the pathogenesis of the infarct.

In the case described, the sequence of events is thought to have been as follows: the patient had chronic vasculitis in the kidney and coronary arteries due to Henoch-Schönlein disease. Vascular endothelial damage caused platelet aggregation which provoked coronary artery spasm and myocardial infarction. Release of vasodilators (such as prostacyclin) led to reversal of the spasm, with distal embolization of the aggregates which were too small to be seen on angiography. Later, systemic emboli from a mural thrombus occurred to the leg and left kidney. The occurrence of this patient's myocardial

infarction, normal coronary infarction, normal coronary arteries and histological evidence of active (renal) vasculitis led the authors to suggest a, possibly, hitherto undescribed myocardial involvement in Henoch-Schönlein disease.

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