Case Report

A fatal case of bowel and cardiac involvement in Henoch–Schoënlein purpura

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Introduction

Henoch–Schoënlein purpura (HSP) is a systemic small vessel vasculitis, characterized by the deposition of immune complexes, mainly polymeric IgA1 and C3, in the skin, gastrointestinal tract, joints, and kidneys [1]. Although the aetiology remains unknown, HSP is often associated with infectious agents. All ages may be affected although it is more common in children, and has a male predominance. Susceptibility to HSP may have a genetic basis. The dominant clinical features of HSP are cutaneous purpura (100%), arthritis (82%), abdominal pain (63%), gastrointestinal bleeding (33%), and nephritis (40%) [2,3]. The overall prognosis is good with a reported mortality of <1% and low morbidity [4]. The case reported here highlights the fatal outcome of an aggressive form of HSP, which had fatal gastrointestinal and cardiac involvement.

Case

A 69-year-old Caucasian male, presented with a 5-week history of ankle pain and bilateral ankle swelling. After taking ibuprofen he developed upper abdominal discomfort and diarrhoea, which did not resolve despite discontinuation of the drug. Three weeks prior to admission, the patient developed an upper respiratory infection with a rash on his legs initially limited to below the level of his knees, which worsened 2 days prior to admission. Past medical history included ischaemic heart disease and duodenal ulcer disease treated surgically. Pre-admission medication included isosorbide mononitrate, aspirin, and simvastatin. He smoked 10 cigarettes/day and had minimal alcohol intake.

At the time of admission he was afebrile, there was no evidence of pallor, cyanosis, jaundice, splinter haemorrhages, or lymphadenopathy. Oral hygiene was poor. He had dark red, non-tender, purpuric lesions on each leg and torso measuring up to 1 cm in diameter and approximately six in number. He was in sinus rhythm, his blood pressure was 140/80 mmHg, heart sounds were normal with no added sounds, peripheral pulses were present and he had 1+ ankle oedema. Auscultation of his chest was normal. Abdominal examination revealed diffuse tenderness most marked in the epigastrium with no rebound tenderness and normal bowel sounds. Initial investigations revealed: haemoglobin 12.4 g/dl, white blood cell count 10.7×109/l, platelets 211×109/l, sodium 134 mmol/l, potassium 4.7 mmol/l, urea 36 mg/dl, creatinine 1.35 mg/dl, random glucose 90 mg/dl, corrected calcium 8.18 mg/dl, bilirubin 0.64 mg/dl, aspartate aminotransferase 15 IU/l, alkaline phosphatase 66 IU/l, serum amylase 47 U/l, albumin 27 g/l, C-reactive protein (CRP) 30.1 mg/l, prothrombin time 18 s (11–18), and partial prothrombin time 25.4 s (26–37). Urinalysis demonstrated haematuria and mid-stream urine culture was negative. Chest X-ray and electrocardiograph (ECG) were both normal.

He deteriorated 48 h after admission with increasing abdominal pain, a small amount of fresh rectal bleeding, and deterioration in renal function (urea 74 mg/dl and creatinine 3.1 mg/dl) despite fluid resuscitation. He was transferred to the regional nephrology unit 4 days after admission. At the time of transfer he was afebrile, his blood pressure was 160/70 mmHg, heart sounds were normal, he had 1+ of bilateral ankle oedema and respiratory bi-basal crepitations, and his abdomen was diffusely tender especially in the left upper quadrant. Investigations on transfer included: haemoglobin
13 g/dl, white blood cell count 12.9 × 10^9/l, platelets 354 × 10^9/l, sodium 130 mmol/l, potassium 5.2 mmol/l, urea 67 mg/dl, creatinine 2.6 mg/dl, corrected calcium 9.4 mg/dl, bilirubin 0.64 mg/dl, aspartate aminotransferase 15 IU/l, alkaline phosphatase 67 IU/l, albumin 22 g/l, amylase 52 U/l, and CRP 122 mg/l. An ultrasound scan demonstrated a mildly enlarged spleen, normal liver, normal size, non-obstructed kidneys with a small amount of free fluid in the abdomen.

The possibility of subacute bacterial endocarditis based on the atypical nature of the rash and the slightly enlarged spleen was also considered and i.v. benzyl penicillin, flucloxacillin, and gentamicin were commenced. CRP decreased and renal function remained unchanged over the next 48 h. Blood cultures were negative and autoantibodies including ANCA, serum immunoglobulins, and complement were all normal. Subsequently, his clinical condition deteriorated with worsening abdominal pain, nausea and retching, inflammatory markers increased, and renal function declined further. An endoscopy was performed which demonstrated purpura at the lower end of the oesophagus. Biopsies were taken. The following day a renal biopsy was performed. The patient continued to deteriorate. There was no radiological evidence of viscus perforation and serum amylase remained normal. An abdominal CT scan demonstrated changes in the small bowel architecture suggestive of either an inflammatory or ischaemic process. At this juncture the renal biopsy result became available. This demonstrated a diffuse endocapillary proliferative glomerulonephritis with 20% crescents plus diffuse staining of the mesangium and capillary walls with IgA, C3, and fibrinogen. This confirmed the diagnosis as HSP (Figures 1 and 2). The oesophageal biopsy was also confirmatory demonstrating a leucocytoclastic vasculitis. The patient was commenced on i.v. methylprednisolone. He continued to remain haemodynamically unstable requiring volume support. The following day he developed biochemical evidence of myocardial damage with elevated CK at 1381 U/l (55–170) and CK-MB at 9.3%. ECG demonstrated left bundle branch block, which was a new finding. He had an EMD arrest 24 h later from which he was successfully resuscitated and transferred to the intensive care unit for ventilation, inotropic support, and haemofiltration. He died later that day, a total of 11 days since initial admission.

Post-mortem examination revealed an infarcted large segment (45 cm) of ileum in the absence of perforation. The histology demonstrated a severe leucocytoclastic vasculitis. Cardiac involvement was limited macroscopically to a 10 mm diameter haemorrhagic lesion localized to the wall of the left ventricle in proximity to the posterior cusp of the mitral valve. Histology of this lesion demonstrated a subendocardial leucocytoclastic vasculitis. There was no evidence of acute myocardial infarction.

Discussion

Although the first description of a case of HSP is attributed to Heberden (1801), it was Schönlein in 1837 who named the association of joint pain and purpura as purpura rheumatica. Later Henoch, a pupil of Schönlein, described the gastrointestinal (1874) and renal involvement (1899) to complete the syndrome. The term anaphylactoid purpura was first used by Frank because the disease was thought to have an allergic aetiology; however, this term is no longer used as this aetiology has not been substantiated. The aetiology of HSP still remains unknown although there have been associations with infectious agents especially group A β-haemolytic streptococci and less commonly measles, rubella, adenovirus, parvovirus, and mycoplasma [5]. The incidence of HSP is highest in the winter months. HSP has been reported in all age groups although by far the most common group affected are children <10 years old. A slight male predominance exists 1.4:1. The majority of affected
individuals recover completely. The extent and nature of renal involvement usually determines the long-term prognosis [6].

Gastrointestinal involvement covers the spectrum from trivial to severe. Endoscopic findings of lower oesophageal purpuric lesions although typical are of no prognostic significance. Occasionally, the abdominal symptoms may mimic an acute surgical abdomen. Major abdominal complications occur in 4.6% of cases (range 1.3–13.6%), of which intussusception is by far the most common in children. In adults, bowel ischaemia, infarction, intestinal perforation, fistula formation, late ileal stricture, acute appendicitis, upper gastrointestinal haemorrhage, and pancreatitis have all been seen although infrequently [7,8]. The extensive small bowel infarction in our patient was an uncommon presentation of HSP, which was contributory to the fatal outcome. Cardiac involvement is very uncommon with documented involvement in HSP being limited to only a handful of case reports in the literature [9–11]. Usually renal involvement may cover a wide spectrum and such involvement is usually regarded as an important long-term prognostic factor [1]. In the case presented here there was diffuse involvement of the glomeruli with 20% crescents (pathological classification HSP IIIb). Renal function survival in adults with HSP-related nephritis is good and has been shown to be comparable at 5 and 10 years, with 85 and 95% of adults and children, respectively, having renal survival. The value at 10 years for both groups is 75% [12]. Nevertheless, these figures tend to mask the fact that HSP in adults represents a more severe clinical syndrome, with a greater frequency of renal involvement. Although the final outcome of HSP is equally good in children and adults, adults invariably require more immunosuppressive treatment [13].

The events leading to the pre-terminal event in this patient, namely the EMD arrest, were most likely a combination of the haemodynamic consequences of severe bowel wall vasculitis together with vasculitis involvement of the conduction system. The fatal outcome in this case highlights the potential severity of HSP and the need to consider early aggressive combined immunosuppressive therapy for certain types of end organ involvement.

References


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