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Pulmonary haemorrhage in Henoch-Schönlein purpura

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ABSTRACT A case of Henoch-Schönlein purpura with fatal pulmonary haemorrhage and capillaritis is described.

Henoch-Schönlein purpura is a syndrome characterised by a rash, arthritis, gastrointestinal manifestations, and nephritis secondary to diffuse necrotising vasculitis. Clinically evident pulmonary disease is very rare and most reports on series of patients do not mention it. We report a patient with fatal intrapulmonary haemorrhage and briefly review the relevant publications.

Case report

A 78 year old man was admitted with a one day history of passing dark red blood rectally. Three weeks previously he had been investigated elsewhere for weight loss, arthralgia, and haemoptysis. Chest radiography had shown patchy opacities in both lower lobes. Blood urea and creatinine concentrations were normal. Sputum culture had been negative, and bronchoscopy had showed an inflamed mucosa. He was treated with penicillin on the presumptive diagnosis of bacterial pneumonia and the haemoptysis had resolved.

On admission he was shocked, and there was a purpuric rash on the buttocks and lower trunk. The blood urea was 50 mmol/l, potassium 7·4 mol/l, and haemoglobin 16·5 g/dl; the platelet count was normal. Clotting studies showed a prothrombin time of 25 (control 10–14) seconds and an activated partial thromboplastin time of 52 (control 30–45) seconds. A chest radiograph was normal.

The patient's condition improved rapidly after resuscitation with intravenous fluids, and the potassium concentration fell to normal values, though the urea remained raised. Oesophagogastroscopy showed haemorrhagic ulcerated oesophagitis. Vitamin K was given and clotting studies gave normal results by the next day.

He remained clinically stable until the third day after admission, when, over minutes, he became acutely dyspnoeic. Chest examination revealed right basal crepitations. Antibiotics were started, but over the next hour he became increasingly breathless and died.

At necropsy the lungs were heavy, oedematous, and congested. Microscopically there was extensive intra-alveolar haemorrhage with fluffy foci of fibrin; hyaline membranes were absent. Capillaritis was present, with loss of nuclear staining in alveolar walls and septa, and capillary wall necrosis (fig 1). Haemosiderin laden macrophages were not seen. The wall of one small artery was infiltrated by neutrophils (fig 2). Immunohistochemical examination showed non-specific staining for IgG, IgM, and IgA in the intra-alveolar fibrin and alveolar walls, but no arterial staining. There was a proliferative glomerulonephritis with crescent formation and increased mesangial matrix, thrombi in some arterioles, and foci of interstitial haemorrhage; immunoglobulin staining was negative.

Discussion

This patient had the classical features of Henoch-Schölein purpura, including a purpuric rash, arthralgia, gastrointestinal bleeding, and glomerulonephritis. At initial presenta-

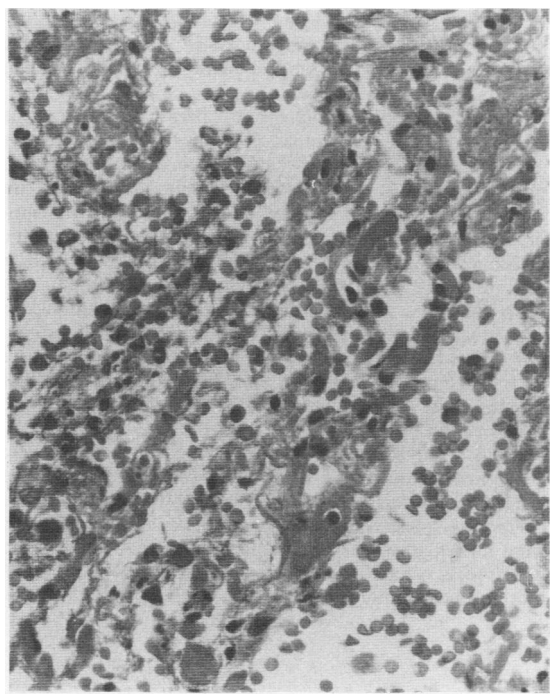

Fig 1 *Lung parenchyma showing necrosis of capillaries and intrapulmonary haemorrhage.*

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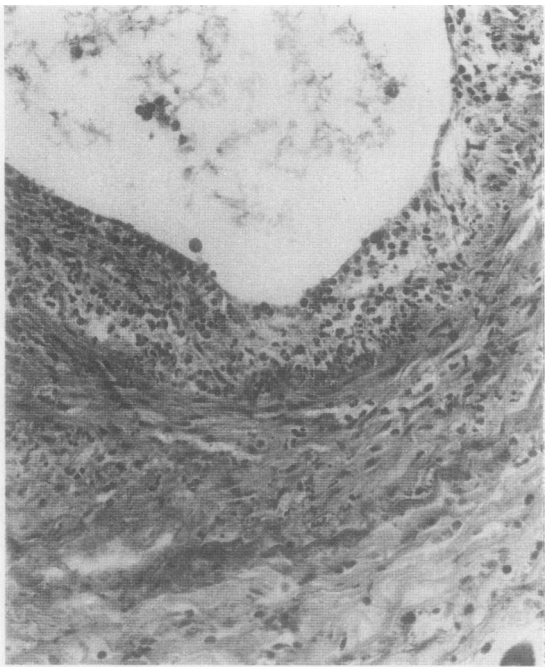

Fig 2 *A small elastic pulmonary artery showing a neutrophilic infiltrate.*

tion there was minor haemoptysis with radiographic opacities, but both had cleared before a final fatal acute pulmonary haemorrhage.

Clinically important pulmonary disease is rare in this condition. It may present as pulmonary haemorrhage or infarction secondary to vasculitis. Transient radiographic opacities are the most common manifestation, and are usually asymptomatic but may be accompanied by haemoptysis. Sometimes there is haemoptysis with no radiographic or bronchoscopic abnormality.[1] Pleuritic chest pain with or without a pleural rub or haemoptysis may occur, leading to a mistaken diagnosis of pneumonia or pulmonary embolus. In such cases anticoagulation will lead to increased bleeding.[1] Even non-fatal pulmonary complications, however, are unusual. In the only large adult series four of 77 patients had symptomatic respiratory disease—haemoptysis, pleuritic pain, or both.[1]

Fatal respiratory complications have been reported on four previous occasions. In three cases there was massive intrapulmonary haemorrhage presenting with dyspnoea and haemoptysis, and resulting in death in one to 48 hours; two were misdiagnosed as pneumonia. These patients all had widespread vasculitis and acute pneumonitis with a polymorphonuclear infiltrate.[2-4] In two there was small vessel leucocytoclastic vasculitis,[3 4] and in the third haemorrhage and pneumonitis obscured the small vessels; the larger vessels were normal.[2] The fourth case had a different presentation. An 8 year old girl with recurrent acute respiratory failure and changing radiographic opacities died after 18 months despite treatment with corticosteroids and cyclophosphamide. Necropsy showed pulmonary alveolar oedema, interstitial fibrosis, and arterial thrombi, but no haemorrhage.[5]

Although one artery showed a neutrophilic infiltrate, the predominant finding in our patient was capillaritis with intra-alveolar haemorrhage. This picture has also been reported in systemic lupus erythematosus and other vasculitides, where the vasculitis usually affects small muscular arteries and arterioles.[6] Pulmonary capillaritis may be more frequent in systemic vasculitis than is generally recognised. Mark and Ramirez[7] studied 13 patients with unexplained diffuse pulmonary haemorrhage with pathologically proved extrapulmonary vasculitis, all of whom had evidence of pulmonary capillaritis. Direct immunofluoresence testing performed in eight cases was non-specific. Pulmonary capillaritis has not been reported in Henoch-Schönlein purpura—although, according to the pathological criteria of Mark and Ramirez, it was present in one of the cases discussed above.[4]

The pathological lesion in Henoch-Schönlein purpura is a widespread vasculitis, IgA probably having a pathogenetic role. IgA deposits have been found in the mesangium and dermal vessels, and circulating IgA complexes have been found early in the disease.[8 9] Pathological changes in the lung have been observed only in postmortem specimens. In one case with pulmonary haemorrhage, leucocytoclastic vasculitis, and probable capillaritis there was extensive granular deposition of IgA along the alveolar septa.[4]

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