Pulmonary Hemorrhage Associated with Henoch-Schönlein Purpura in Pediatric Patients: Case Report and Review of the Literature

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Objectives: To characterize the clinical characteristics of pediatric patients with pulmonary hemorrhage and Henoch-Schönlein purpura (HSP).

Methods: Presentation of a pediatric patient with pulmonary hemorrhage associated with HSP and review of relevant cases based on a PubMed search (1966 to April 2010).

Results: We identified 17 previously reported pediatric cases with HSP and pulmonary hemorrhage. The most frequent clinical manifestations were rash (17 patients, 100%), proteinuria (14 patients, 82%), and abdominal pain (13 patients, 76%). Six patients (35%) had complete resolution of symptoms; 7 patients (41%) had partial recovery, and 4 patients (23%) died. Nine patients (53%) had acute respiratory failure following intubation and 3 of these patients (33%) died. Five patients were treated with methylprednisolone pulse therapy and 1 was also given low-dose cyclophosphamide treatment, but 2 of these 5 patients (40%) died. Three patients were given cyclophosphamide pulse therapy plus steroid (nonpulse or pulse) therapy, and all survived. Among the 6 nonintubated patients, all were given steroid treatment with or without an immunosuppressant drug, and all survived. In our reported case, plasma exchange appeared to help resolve the pulmonary hemorrhage and crescentic glomerulonephritis that were associated with HSP.

Conclusions: For pediatric HSP patients with pulmonary hemorrhage but no respiratory failure, methylprednisolone pulse or nonpulse therapy could be the first-line therapy. In the presence of respiratory failure, cyclophosphamide pulse therapy is suggested. Plasma exchange may be considered for treatment of pulmonary renal syndrome or refractory pulmonary hemorrhage.

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Keywords: *Henoch-Schönlein purpura (HSP), pulmonary hemorrhage, pulmonary-renal syndrome, crescentic glomerulonephritis, methylprednisolone pulse therapy, plasma exchange*

Henoch-Schönlein purpura (HSP) is a small-vessel vasculitis characterized by a purpuric rash, arthritis, and abdominal pain (1). Most patients are 4 to 7 years old, but the disease may also appear in infants and adults (2). The prognosis is generally excellent, but severe complications can occur. The most common complication is renal involvement, especially acute nephritis, nephrotic syndrome, and acute renal failure (3,4). Pulmonary hemorrhage is a rare complication of HSP, especially in pediatric patients (5-16), but is associated with high mortality and morbidity (12). We describe a pediatric patient with HSP complicated by pulmonary hemorrhage and crescentic glomerulonephritis and analyze all previously reported cases of pediatric pulmonary hemorrhage associated with HSP.

METHODS

We present a previously unreported case of pediatric HSP with pulmonary hemorrhage. A PubMed (www.ncbi.

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The authors have no conflicts of interest to disclose.

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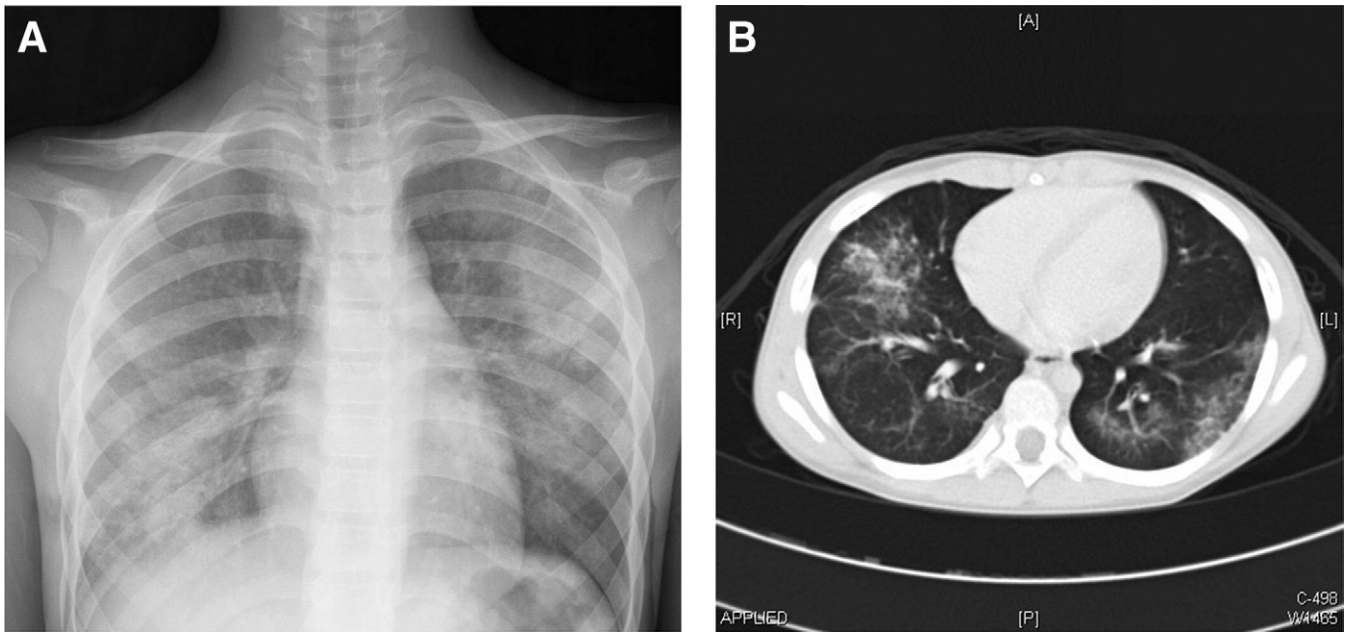

Figure 1 Evidence of pulmonary hemorrhage. A, Chest radiograph showing bilateral diffuse consolidation; B, Chest CT scan showing multiple ground-glass opacities.

nlm.nih.gov/pubmed) search of the literature (1966 to April 2010) was performed to identify all previously reported cases of pulmonary hemorrhage associated with HSP by using the terms Henoch-Schönlein purpura, pediatric, pulmonary hemorrhage, and pulmonary renal syndrome. We found 16 cases and summarized the reported symptoms, clinical findings, treatments, and outcomes.

CASE REPORT

In March of 2009, an 11-year-old girl, who had HSP diagnosed by intermittent abdominal pain, skin rash, and small intestine biopsy when she was 4 years old, was referred to our emergency department due to intermittent abdominal pain and multiple purpura over her elbows and lower legs for the previous 3 days. She also had a mild cough without sputum, vomiting, anorexia, and general malaise. A physical examination indicated multiple purpura over her elbows and distal legs and bilateral coarse breath sounds without crackle or wheeze.

Serum analysis indicated the following: white blood cell count, 11,500/mm^3; hemoglobin, 6.9 g/dL; platelet count, 690 $\times$ 10^3/mm^3; urea, 20.0 mg/dL; creatinine, 1.02 mg/dL; C-reactive protein, 18.05 mg/dL; total protein, 6.5 g/dL; albumin, 3.4 g/dL; total cholesterol, 236 mg/dL; D-dimer, 7881.43 ng/mL; and fibrinogen, 553 mg/dL. Urine analysis indicated hematuria and proteinuria. A chest radiograph (CXR) indicated bilat-

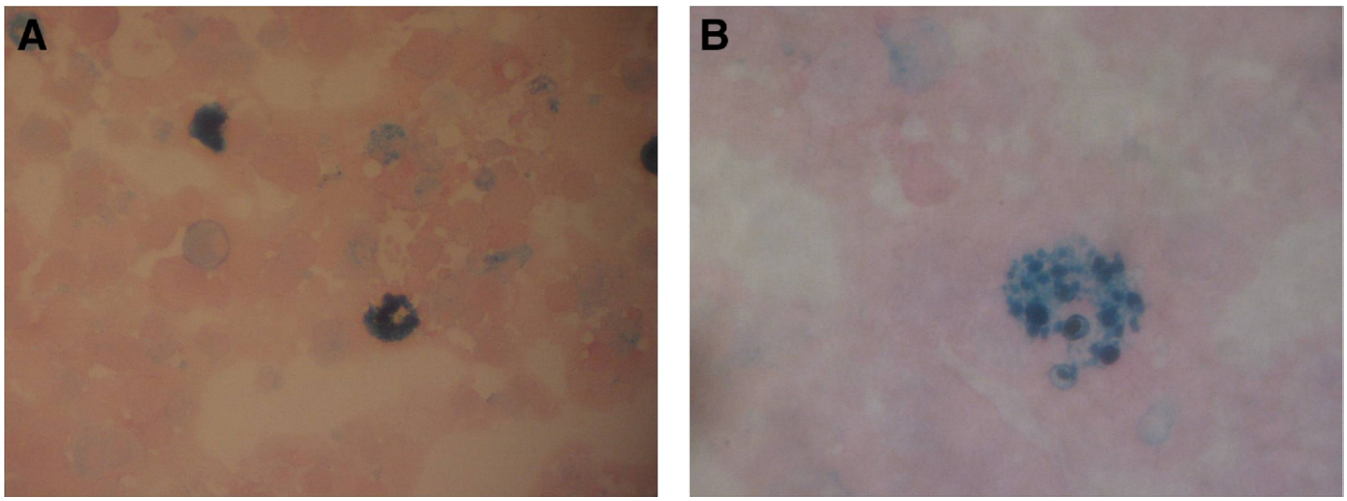

Figure 2 Evidence of hemosiderin-laden macrophages (A and B) in the bronchioalveolar lavage. (Color version of figure is available online.)