A Case of Pulmonary Infarction Probably Related with Henoch-Schönlein Purpura

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We report a case of pulmonary infarction probably related with Henoch-Schönlein purpura, which presented with purpura on both lower extremities in a 27-year-old woman. The purpura had developed 4 days previously and it had no itching or tenderness. She complained of mild fatigue, chest tightness, but no dyspnea or hemoptysis. On routine examination, chest radiography showed a poorly defined nodular opacity on the left lung field, and it was diagnosed by a follow up chest CT as a pulmonary infarction secondary to pulmonary vasculitis.
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Key Words : Henoch-Schönlein purpura, Pulmonary infarction, Pulmonary vasculitis

Henoch-Sch nlein purpura (HSP), also known as anaphylactoid purpura or allergic purpura, is a hypersensitivity reaction in the immunogenetically susceptible host. Immunohistologically it shows immune complex deposition in the walls of affected blood vessels and perivascular areas. HSP is characterized by nonthrombocytopenic purpura accompanied by arthritis or arthralgia, abdominal pain due to gastrointestinal bleeding, and glomerulonephritis[1]. HSP is primarily a disease of children, but can occur at any age. The skin lesion typically involves the extensor part of extremities and buttocks. Pulmonary involvement in this disease is rare and primarily includes hemorrhage and/or interstitial disease[2].

We report a case showing features of a pulmonary infarct in Henoch-Schönlein purpura.

CASE REPORT

A 27-year-old woman attended the Department of Dermatology in May 1996, complaining of palpable purpura on both lower extremities(Fig. 1) and buttocks. It had developed 4 days previously and she had no itching or tenderness. The patient had previously been in good health and had no pertinent medical history. In addition there was no significant family history.

On systemic review, the patient complained of arthralgia, fatigue and mild chest tightness but did not show dyspnea or hemoptysis. Multiple various sized violaceous palpable purpura were shown on the lower extremities and buttocks. Chest tightness developed at the same time with purpura. Laboratory findings were leukocytosis (10,450/mm^3), an elevated erythrocyte sedimentation rate (45mm/hr) and platelet count (534,000/mm^3). Urine analysis showed some increase in epithelial cells, but it returned to normal in the 2 and 4 week follow up examinations. Other findings(anti-nuclear antibody, anti-neutrophil cytoplasmic antibody, C3, C4) were all within normal limits. On radiographic examination, the chest X-ray showed a poorly-defined cavitary nodular opacity (Fig. 2), but she showed no specific findings on the previous chest X-ray taken 1 month previously. We followed up with chest computerized tomography and it revealed a wedge shape consolidation and ground glass attenuation probably due to hemorrhage of

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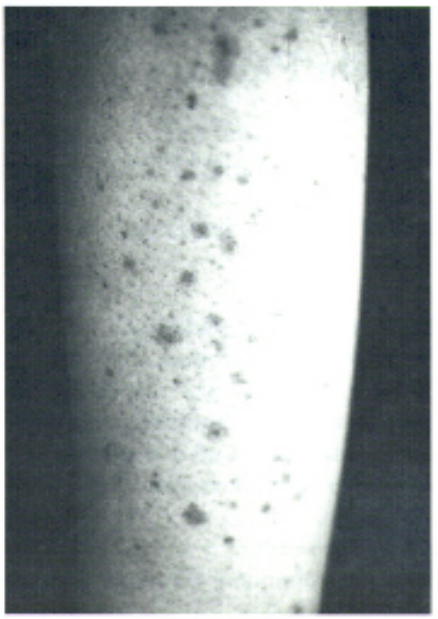

Fig. 1. Multiple pinhead to match-head sized violaceous purpura on the lower extremities.

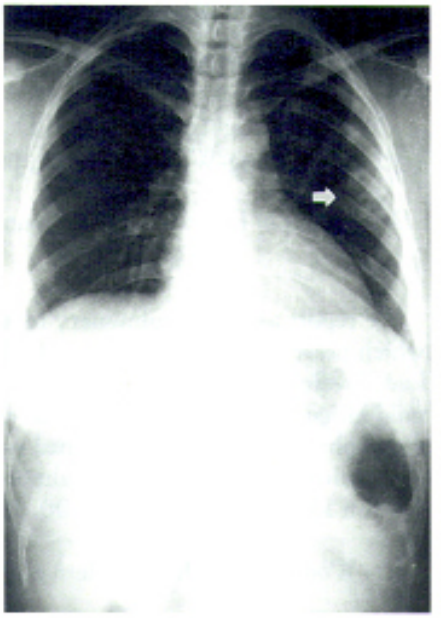

Fig. 2. Multiple pulmonary infarction probably due to pulmonary vasculitis on chest X ray(poorly defined nodular opacity : arrow).

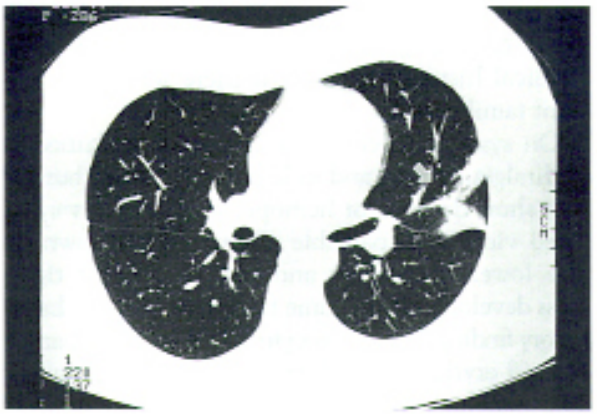

Fig. 3. Pulmonary infarction probably due to pulmonary vasculitis (shows haziness around vessel : arrow).

the anterior and posterior segment of left upper lobe (Fig. 3). Therefore we diagnosed the lesion as pulmonary infarction due to pulmonary vasculitis. The histological finding in the lower leg showed the dermal capillary with swelling of endothelial cells and focal deposition of fibrinoid material around the vessel walls. Severe perivascular inflammatory infiltrates composed of neutrophils were seen (Fig. 4A & 4B). An immunofluorescent study was performed on the specimen, and it showed a deposit of IgA in the blood vessels at the papillary dermis.

Treatment included an intramuscular injection of triamcinolone 40mg, and oral prednisolone 30mg daily for 6 days; An improvement in the skin lesions was seen and the patient did not develop further pulmonary symptoms. Pulmonary radiographic changes resolved on follow up X-ray 3 weeks later. She remained clinically stable during the following 6 months and did not reveal any skin lesions or other complications.

DISCUSSION

Henoch-Schönlein purpura is a kind of hypersensitivity vasculitis syndrome characterized by a non-thrombocytopenic purpura, arthralgia/arthritis, abdominal pain due to gastrointestinal hemorrhage and hematuria due to glomerulonephritis. Clinically pulmonary disease is very rarely present[3]. The onset of the skin lesion is frequently preceeded by constitutional symptoms, including fever, malaise or anorexia for up to several days. The 1990's classification of HSP from the American College of Rheumatology requires at least two of

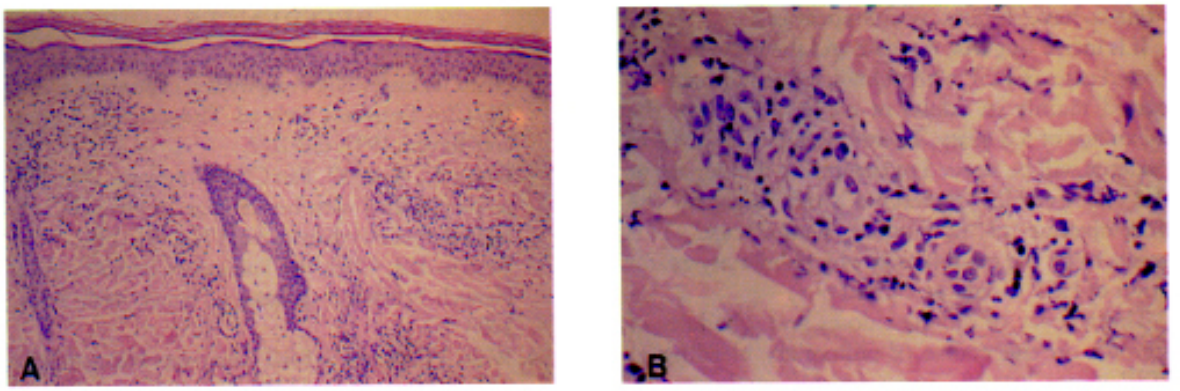

Fig. 4. (A) Upper dermis and perivascular space shows patch infiltration of neutrophils (H & E ×100). (B) Dermal vessels shows endothelial swelling. Severe inflammatory infiltrates of neutrophils around the vessel and neutrophils show fragmentation of nuclei (H & E ×400).

the four following criteria for diagnosis: palpable purpura, age younger than 21 years at onset, bowel angina, and/or a skin biopsy specimen with intramural neutrophils in dermal vessels1. HSP is primarily a disease of childhood, with most cases occurring between the ages of 3 and 10 years[1]. Although atypical cases affecting adults have been reported frequently, patients with classic findings including systemic involvement are usually limited to the pediatric population[1]. Older patients or those with atypical clinical features may require a skin biopsy with or without immunofluorescent staining to establish the correct diagnosis.

It has been reported that respiratory complications of HSP are very rare and transient radiographic opacities are the most common manifestations. But in a series of adult patients, 8 of 141 with HSP were reported to have pulmonary complications[2,3]. Clinically fatal complications are rare and occur as pulmonary hemorrhage or infarction secondary to vasculitis. But most patients with pulmonary complications are asymptomatic and may be accompanied by minor hemoptysis. Sometimes there is hemoptysis with no radiographic or bronchoscopic abnormality. Pleuritic chest pain with or without a pleural rub may occur[4,5]. This condition may lead to a mistaken diagnosis of pneumonia or pulmonary embolus. In such misdiagnosed cases, anticoagulant therapy will lead to increased bleeding. But minimal pulmonary hemorrhage due to HSP can be improved rapidly with other systemic manifestations without complications.

In our case, despite the chest X-ray findings, the patient did not complain of specific symptoms except for mild chest tightness. However, other cases in a series of studies showed that, 3 of 141 adult patients had massive pulmonary hemorrhage, dyspnea, hemoptysis and died after 48 hours[3]. These patients all had widespread vasculitis and inflammatory infiltration. In two cases there was small vessel leukocytoclastic vasculitis, and all patients had common inflammation at small vessels and intraalveolar hemorrhage. It is commonly accepted that the characteristic feature of renal and vascular regions in HSP is the deposition of circulating immune complexs containing IgA. However, 25% of HSP patients did not have IgA immunofluorescence of involved skin. Whereas IgA immunofluorescence would have supported the diagnosis of HSP, its absence does not negate the diagnosis of patients. Alveolar hemorrhage may occur with other vasculitis or autoimmune diseases like Wegener's granulomatosis and systemic lupus erythematosus. But anti-neutrophilic cytoplasmic antibodies, which recently have been associated with Wegener's granulomatosis, were negative in HSP.

The course of HSP is characterized by episodes of remission and relapse, with complete resolution occurring in the majority of patients within 1 to 2 months. Long term prognosis is dependent on the extent of the systemic involvement.

The recurrence of pulmonary complications in HSP is rarely observed. Olson et al reported 4 patients who developed pulmonary hemorrhage in association with HSP, but follow up at 14 to 30 months after the pulmonary hemorrhage revealed

none to have respiratory complications[2].

It is interesting that our case and the previously reported cases of HSP with pulmonary hemorrhage occurred mainly in adults or cases older than the mean age of 5.8 years. Therefore larger numbers of patients will be needed to determine if older age at the onset of HSP represents a risk factor for the development of pulmonary hemorrhage. This seems to be the main reason why pulmonary evaluation of HSP should be carried out[6].

The efficacy of different doses and schedules of steroids on the course of pulmonary bleeding in HSP is not known. Some articles emphasize aggressive treatment of pulmonary hemorrhage with pulse methyl-prednisolone or immunosuppressive agents[2]. Plasmapheresis may improve purpura and renal function by enhanced removal of circulating IgA[7].

In conclusion, a 27-year-old women was seen with a feature of HSP with pulmonary infarction, which improved without severe pulmonary complications during a follow up period of 4 weeks.

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