□ SHORT COMMUNICATION □

“Crazy-paving” Appearance in Systemic Lupus Erythematosus

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A 21-year-old woman was referred to our hospital in April 2003 with a 2-month history of fever and butterfly rash. Laboratory studies on admission were as follows: white blood cell count, $1.3 \times 10^9/L$; hemoglobin, 8.2 g/dL; platelet count, $144 \times 10^9/L$; anti-nuclear antibody titre, 1: 640; anti-DNA antibody, 1,261 U/mL; CH50, <5.0 U/mL; anti-Sm antibody, 17.5 (normal range <7.0) and anti-RNP antibody, 17.5 (normal range <15). Lupus anticoagulant, anti- β_2 GPI antibody, anti-GBM antibody, anti-SS-A antibody, anti-SS-B antibody and MPO-ANCA were negative. Chest roentgenography and renal function on admission were normal. Systemic lupus erythematosus (SLE) was diagnosed and oral prednisolone (40 mg/day) was begun. Fourteen days after starting treatment, the patient developed a non-productive cough (without hemoptysis or hemosputum) and hemoglobin fell to 6.7 g/dL. Chest roentgenography demonstrated reticular shadowing, predominantly in the lower lung fields (Fig. 1A). Thin-section chest CT showed a fine reticular pattern superimposed on a background of ground-glass opacity; i.e. crazy-paving appearance (Fig. 1B) predominantly in the middle to lower lobe of the bilateral lung. Bronchoscopy was performed in order to differentiate between infection and diffuse alveolar hemorrhage (DAH). As bronchoalveolar lavage at the rt. middle lobe demonstrated hemoserous fluid, and no evidence of infection was obtained, a diagnosis of DAH due to SLE was made. Methylprednisolone pulse therapy was therefore added, resulting in improvement of symptoms and radiographic appearance.

The crazy-paving appearance results from a fine linear network superimposed on an area of ground-glass opacity on thin-section CT. This finding has been considered strongly suggestive of alveolar proteinosis (1). However, in our clinical experience and in several reports in the literature, other diseases such as acute interstitial pneumonia, acute respiratory distress syndrome, cardiogenic pulmonary

edema, mucinous bronchoalveolar carcinoma, and lipoid pneumonia can have this appearance on thin-section CT (2-5).

Non-infectious pleuropulmonary complications of SLE occur at some point during the course of the illness in 50%-70% of affected individuals (6). These include pleuritis and pleural effusions; interstitial lung disease such as acute lupus pneumonitis; pulmonary embolism (usually associated with the antiphospholipid syndrome); pulmonary vascular disease including vasculitis and a primary form of pulmonary hypertension; diaphragmatic weakness; and bronchiolitis (6). Although most of these complications have a low mortality rate, DAH is considered a catastrophic and often lethal pulmonary complication of SLE.

Symptoms of DAH are usually acute, developing over hours to several days, although in some cases a more gradual onset is seen. Although the hemoglobin level often drops significantly and diffuse infiltrates on chest radiography are characteristic, hemoptysis is not necessarily present at the time of presentation. No uniform recommendations for treatment have been established; however, corticosteroids remain the mainstay of therapy. Various combinations of immunosuppressive drugs and plasmapheresis have also been recommended.

The underlying histopathological finding in the majority of reported cases is a form of “bland” alveolar hemorrhage without underlying vasculitis, diffuse alveolar damage, or interstitial inflammation, unless accompanied by an infectious pneumonia. Several authors have reported that pulmonary vasculitis, as evidenced by pulmonary capillaritis and microangiitis, is the causative mechanism of DAH in patients with SLE (7).

In a post-mortem study, Johkoh et al (8) described the spectrum of pulmonary diseases associated with crazy-paving appearance on thin-section CT and the underlying pathologic features. They classified the pathophysiological mechanisms behind the fine reticular patterns apparent on pathologic analysis into three types of processes: type 1, an alveolar filling process (i.e., air space disease); type 2, an interstitial fibrotic process; and type 3, a combination of both interstitial and intraalveolar diseases. They stressed that the reticular patterns on the areas of crazy-paving do not necessarily result from thickening of interlobular septa or the presence of intralobular fibrosis, but can also be caused purely by airspace disease. In this case, the appearance seems to be a consequence of linear deposition of material within the airspaces.

As Johkoh et al (8) reported the prevalence of the crazy-

Figure 1. Chest roentgenogram (A) showing reticular shadow predominantly in bilateral lower lung fields, and high resolution CT of the lung (B) showing a fine reticular pattern superimposed on a background of ground-glass opacity.

paving appearance in pulmonary hemorrhage was 9%, it is important to remember DAH. However, little has been known about the crazy-paving appearance presented in patients with DAH complicating SLE. In conclusion, we con-

sider that the crazy-paving pattern on thin-section CT scans is a nonspecific finding and it can be seen even in patients with DAH complicating SLE.

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