THE CRAZY–PAVING PATTERN; A NONSPECIFIC SIGN ON HRCT CHEST

Piyaporn Limanond; M.D., Orasa Chawalparit; M.D., Trongtum Tongdee; M.D., Poonsook Jitnuson; M.D., Pornpim Fuangtharntip; M.D. and Kobkun Muangsomboon; M.D.

ABSTRACT

Purpose : To report the crazy–paving pattern on High Resolution Computed Tomography (HRCT) chest found in our institute which were caused by various processes other than pulmonary alveolar proteinosis (PAP)

Materials and Methods : From June 1998 to August 1999, 4 patients (3 male and 1 female, mean age 38.5 years, range 23-52 years) with crazy-paving pattern on HRCT chest were diagnosed by 2 radiologists. Retrospective review of the imagings was performed with clinical and pathologic correlation.

Results : The crazy-paving pattern in our institute was caused by various processes, including 1 bronchioloalveolar carcinoma, 1 lipoid pneumonia, 1 PAP and 1 pulmonary hemorrhage.

Conclusion : From our findings, we can conclude that the crazy–paving pattern is only a nonspecific sign on HRCT chest, resulted from multiple causes. Clinical and pathologic correlation are necessary for differential diagnosis.

INTRODUCTION

The crazy–paving pattern on High Resolution Computed Tomography (HRCT) chest was previously known as a characteristic sign of pulmonary alveolar proteinosis (PAP), consisting of geographic areas of ground-glass alveolar infiltration with superimposed smooth thickening of interlobular septa.

Based on open lung biopsy, well-demarcated geographic areas of ground-glass consolidation are explained by filling of alveolar spaces with PAS-positive phospholipoprotein materials whereas superimposed smooth interstitial thickening reflects septal edema and alveolar wall infiltration by lymphocytes and macrophages.

We report 4 cases of crazy-paving pattern on HRCT chest; found in our institute with clinical and pathologic proving not to be caused by only PAP but various causes.

MATERIALS AND METHODS

From June 1998 to August 1999, 4 patients (3 male and 1 female); age range from 23 – 52 years (mean age 38.5 years); showed crazy-paving pattern on HRCT chest. The imagings were blindly reported by 2 radiologists without patho-

Department of Radiology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, THAILAND

logic correlation.

The HRCT chest of these patients were performed by Philips Tomoscan AVE 1, using 1-mm collimation, high-spatial frequency reconstruction algorithm, 1-second scan time, 120-140 kVp, 150-175 mA and 512x512 matrix size.

Retrospective review with clinical and pathologic proving of these 4 patients were done, showing variation of causes.

RESULTS

Case 1 : a 33-year-old man presented to his primary physician with chronic cough and blood-tinged sputum. A chest radiograph showed minimal fibronodular infiltration at RUL, that was diagnosed to be pulmonary tuberculous infection. He had received anti-TB drugs for 6 months but no clinical improvement. A 1-year subsequent chest radiograph revealed progression of RUL infiltration with an irregular-walled cavity at RUL. The HRCT chest (Fig. 1) demonstrated a geographic area of ground-glass consolidation at the posterior segment of RUL with superimposed smooth thickening of interlobular septa (crazy-paving pattern). A large cavity about 5 x5 cm was demonstrated nearby the consolidation. It had thick irregular wall with internal septation. Bronchoscope and transbronchial biopsy showed bronchogenic carcinoma, non-small cell type.

Case 2 : A 23-year-old man presented with productive cough and progressive dyspnea for 2 months. A chest radiograph showed bilateral alveolar infiltration. Subsequent HRCT chest (Fig. 2) revealed diffuse ground-glass opacity with smooth interlobular septal thickening (crazy-paving pattern) in both lung fields. The pathologic findings from lung biosy highly suggested lipoid pneumonia with interstitial fibrosis.

Case 3 : A 52-year-old man presented with dyspnea for 1 month, showing bilateral interstitial infiltration in chest radiograph. The HRCT chest (Fig. 3) demonstrated discrete geographic ground-glass consolidation with superimposed interlobular septal thickening (crazy-paving pattern) in both lung fields, a highly suggestive sign for PAP. Bronchoscopic and bronchoalveolar larvage findings also supported the diagnosis.

Case 4 : A 46-year-old woman presented to her chest physician with cough and blood-tinged sputum for 3-4 days. No history of weight loss or previous trauma was interviewed. Her chest radiograph showed normal findings but subsequent HRCT chest (Fig. 4) revealed a small area of crazy-paving pattern at the anteromedial basal segment of LLL. Her first bronchoscope revealed an organized hematoma at the basal segment of LLL bronchus without cytologic or histologic evidence of malignancy. Her symptoms were spontaneously improved without any treatment. A 1-week-subsequent bronchoscope showed resolving hematoma with nearby dilated bronchial vessels. Her chest physician believed that the crazy-paving pattern was due to pulmonary hemorrhage from spontaneous rupture of dilated bronchial vessels.

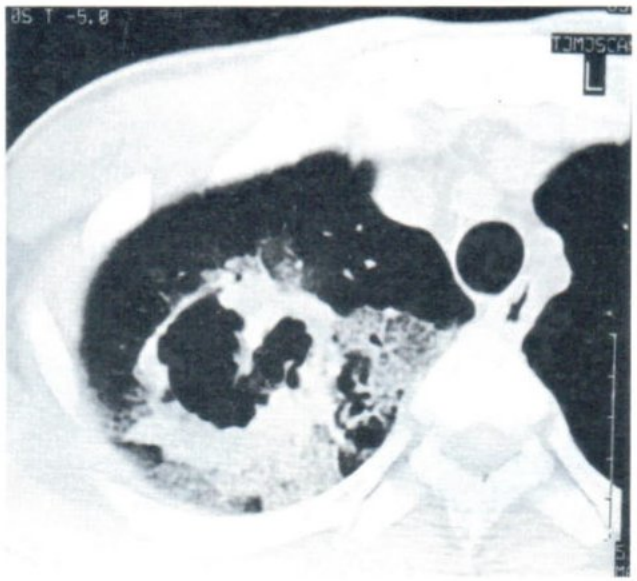

Fig. 1 Bronchogenic carcinoma, non-small cell type. HRCT chest of a 33-year-old man, who presented with chronic cough and blood-tinged sputum, revealed the crazy-paving pattern at the posterior segment of RUL with a nearby large cavity showing thick irregular wall and internal septation. Transbronchial biopsy showed bron chogenic carcinoma, non-small cell type.

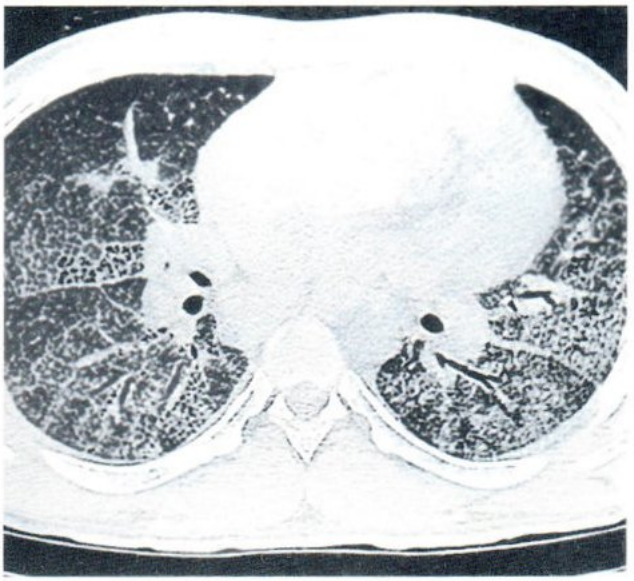

Fig. 2 Endogeneous lipoid pneumonia. HRCT chest of a 23-year-old man, who presented with productive cough and progressive dyspnea for 2 mo., revealed diffuse crazy-paving pattern in both lungs fields. The pathologic findings highly suggested to be lipoid pneumonia.

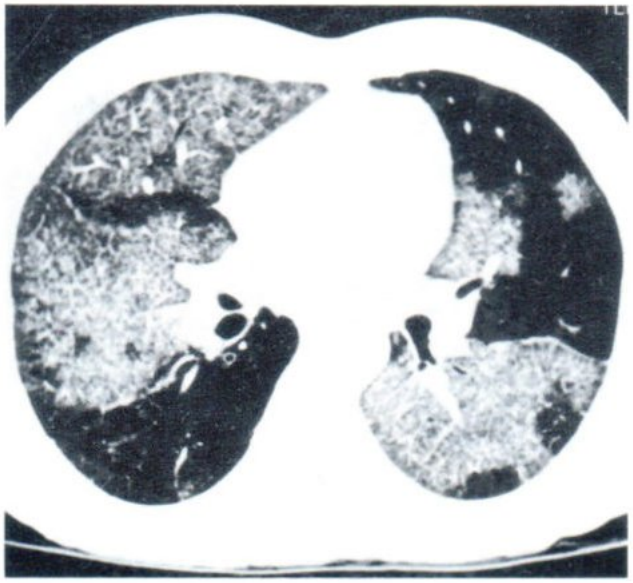

Fig. 3 PAP. HRCT chest of a 52-year-old man, who presented with dyspnea for 1 mo., demonstrated discrete geographic areas of crazy-paving pattern in both lung fields. Pathologic findings supported the diagnosis of PAP.

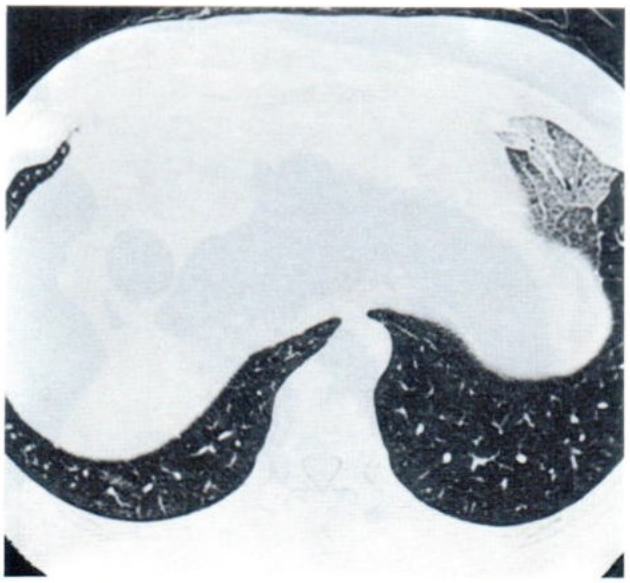

Fig. 4 Pulmonary hemorrhage. HRCT chest of a 46-year-old woman, who presented with cough and blood-tinged sputum for 3-4 days and had spontaneous improvement without any treatment, demonstrated a small area of crazy-paving pattern at the anteromedial basal segment of LLL. Two bronchoscopic studies showed an organized hematoma at the basal segment of LLL bronchus which rapidly dissappeared in 1 wk. and nearby dilated bronchial vessels.

DISCUSSION

Pulmonary alveolar proteinosis (PAP) is an idiopathic disease which associates with dust exposure (silicoproteinosis) and other immunodeficiency. Its pathophysiology is overproduction of surfactant by pneumocyte type 2 with defective clearance of surfactant by alveolar macrophage. Surfactant-filling alveoli cause sharp-demarcated geographic areas of ground-glass consolidation on HRCT chest. The typical pattern of alveolar infiltration in pulmonary alveolar proteinosis is rather symmetrical ground-glass pattern in bilateral lung fields, possibly diffuse or bilateral perihilar areas (bat-wing appearance) with tendency to be prominent at lung bases. Superimposed smooth thickening of interlobular septa are pathologically proved to be septal edema and alveolar wall infiltration by inflammatory cells. Ground-glass consolidation and superimposed interlobular septal thickening cause a well known radiologic sign on HRCT chest called Crazy-Paving Pattern which was previously known as a characteristic sign for PAP. Although Webb et al.[1] suggested that the crazy-paving pattern can be found in other diseases such as pneumocystis and cytomegalovirus infection; until now there is a little information about other causes.

Previously, Tan and Kuzo[3] reported the crazy-paving pattern in a patient with bronchioloalveolar carcinoma; a well differentiated primary lung cancer that associates with preexisting pulmonary scar and fibrosis. Bronchioloalveolar carcinoma arises along peripheral bronchiolar and alveolar walls with absence of an intrinsic bronchial tumor. It has a tendency to spread locally; using lung structure as a stroma (lepedic growth) without disrupting lung architecture. Bronchioloalveolar carcinoma has various radiologic and CT findings such as a single mass, multiple nodules or diffuse (pneumonic) form. Crazy-paving pattern is also one of its findings. Ground-glass opacity reflects alveolar filling of glycoprotein materials from mucous-producing cells. Superimposed interlobar septal thickening represents lepedic growth of tumor cells or lymphatic edema.

Franquet et al.[4] recently reported the crazy-paving pattern in 3 patients with exogeneous lipoid pneumonia. They had a history of aspiration or inhalation of oily substance and subsequent pulmonary inflammation. The CT-pathologic correlation showed that areas of ground-glass consolidation with negative Hounsfield units represented alveolar filling with oily substance and lipid-laden macrophages. Interlobular septal thickening reflected interstitial infiltration with inflammatory cells and variable amount of fibrosis. Due to aspiration cause, the common areas of pulmonary involvement in exogeneous lipoid pneumonia are gravity-dependent areas such as RML and lower lobes. On the contrary, endogeneous lipoid pneumonia has an unknown etiology and not associates with history of external oil used. Pulmonary involvement of endogeneous lipoid pneumonia are also not gravity-dependent areas. Our patient (case 2) had no history of external oil used and showed diffuse crazy-paving pattern in his HRCT chest, compatible with endogeneous lipoid pneumonia.

In case of pulmonary hemorrhage; ground-glass opacity reflected alveolar filling by hemolyzed blood. Interlobular septa in area of consolidation appear thicker and denser than normal, explained by excellent contrast resolution of pulmonary interstitium stand out prominently from the background of low-density material filled alveoli.

We report 4 cases of patients with crazy-paving pattern in HRCT chest, found in our institute which have a variation of causes (1 non-small cell lung cancer, 1 lipoid pneumonia, 1 PAP and 1 pulmonary hemorrhage). Although a small number of patients were included in this study, we can prove that the crazy-paving pattern is only a nonspecific finding which can be found in various processes. Ground-glass opacity represents alveolar filling of low density materials (PAS-positive phospholipoprotein materials in PAP, glycoprotein in bronchioloalveolar carcinoma, oily substance in lipoid pneumonia and hemolyzed blood in pulmonary hemorrhage). Superimposed smooth interlobular septal thickening reflects tumor or inflammatory cell infiltration, septal edema and fibrosis. So pathologic correlation is sometimes important for definite diagnosis. However, there are some clues which can be helpful for differentiation such as previous history, distribution of pulmonary involvement, other associated CT findings and the progression of disease.

Pulmonary alveolar proteinosis (PAP) associates with history of dust exposure (silicoproteinosis) or other immunodeficiency. It has a gradual onset of symptoms with a slow progression of disease. The pattern of ground-glass opacity in PAP are usually symmetrical which is helpful in the differential diagnosis from other diseases. It is common to involve bilateral perihilar areas, prominent at lung bases.

Bronchogenic carcinoma is suspected in elderly patient with preexisting pulmonary scar or fibrosis. Asymmetrical involvement, associated lymph nodes and pleural effusion are helpful in the differentiation from PAP.

Exogeneous lipoid pneumonia is highly suspicious in patient who has a history of aspiration or inhalation of oily substance, contrasting with endogeneous lipoid pneumonia. Clinical course of lipoid pneumonia has a gradual onset and a slow progression like PAP. Exogeneous lipoid pneumonia usually involves asymmetrical gravity-dependent areas such as RML and lower lobes. However endogeneous lipoid pneumonia such as our patient (case 2) was different. He had diffuse ground-glass opacity in both lung fields. Lipoid pneumonia sometimes demonstrates areas of low-attenuated consolidation with negative Hounsfield units (oily substance). However due to reactive inflammation and subsequent fibrosis, oily substance is sometimes obscure and so did our patient.

A remarkable feature of pulmonary hemorrhage is rapid resolution. It sometimes enlarges in early 48-72 hours after its onset (possible having a history of previous trauma) and usually has complete resolution in 2 – 10 days.

CONCLUSION

Crazy-paving pattern is an infrequent finding on HRCT chest. Previously, it was reported as a characteristic finding in pulmonary alveolar proteinosis (PAP). However the more HRCT chest is performed; the more crazy-paving pattern is found. Now it is no longer being a specific sign for PAP but has a variation of causes. Other than pathologic correlation; clinical history, distribution of pulmonary involvement and associated CT findings are necessary for a definite diagnosis.

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