Collation Of Fibrocystic Pathology With Constant Respiratory Illness

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Abstract

Objectives: This retrospective study examines the differences of the fibrocystic stage of Pathology with idiopathic interstitial fibrosis using High-resolution Computed Tomography (HRCT), imaging features and quantification of the severity and extent of the disease.

Methods: 81 patients with biopsy diagnosed pulmonary sarcoidosis and 81 patients with Constant interstitial pneumonia (UIP) were included. All underwent HRCT scan. Findings were evaluated per lung lobe. The severity of disease was quantified per feature and overall with the Gay-Watters score.

Conclusions: The micro nodular pattern highly indicates Pathology, whereas honeycombing and ground glass with traction bronchiectasis are associated with UIP. These features can assist on differentiation, when disease presents itself with Fibrocystic HRCT characteristics at diagnosis.

Keywords: Fibrocystic Pathology, Constant interstitial pneumonia, HRCT

Introduction

Sarcoidosis, a multisystem chronic inflammatory condition of unknown etiology, is characterized by nongaseous epithelioid cell granulomas and changes in tissue architecture. The mediastinal and Hilary lymph nodes enlargement is the most common imaging feature in the chest, presented in approximately 80-90% of the patients

[1]. The most common pulmonary symptoms are cough, dyspnea and chest pain. Other symptoms include fatigue, night sweats, weight loss, and erythema nodosum. 50% of the reported cases is asymptomatic, with abnormalities detected incidentally at chest radiography

[2]. A diagnosis of Pathology is established on the basis of compatible clinical-radiological findings and histological evidence of the presence of nongaseous epithelioid cell granulomas in one or more organs. High-resolution computed tomography (HRCT) provides detailed information regarding lung involvement and hence can be helpful for distinguishing active inflammation from irreversible fibrosis in selected patients with stage 2 or 3 Pathology

[3]. Nodules, ground-glass opacities, and alveolar opacities are suggestive of granulomatous inflammation that may be reversed with therapy, whereas honeycomb pattern, architectural distortion, volume loss, and traction bronchiectasis are indicative of irreversible fibrosis

[4]. In fact, Fibrocystic interstitial disease is a common pattern at presentation. HRCT may also be useful for verifying specific diagnoses in patients with atypical clinical manifestations or un Constant radio graphical features

[5]. Constant interstitial pneumonitis, or idiopathic pulmonary fibrosis (UIP/IPF), is a common type of interstitial pneumonia with progressive diffuse parenchymal lung changes and unclear pathophysiology

[6]. The median survival is poor, recording 2-3 years

[7]. Restrictive diseases, such as UIP/IPF, result in decreased lung volumes and distortion of the normal anatomy. In diffuse lung diseases, the HRCT is used as a significant tool. Several studies have shown that HRCT of the chest with certain features can be used as a surrogate in order to recognize the pathological pattern of UIP with high degrees of confidence

[8]. In this context, the present study investigates the differences between the Fibrocystic stage of Pathology and fibrosis in UIP/IPF using High-resolution Computed Tomography by comparing individually the imaging features, the severity and the extent of the disease of each entity. This may be of assistance in imaging differentiation between the two entities, since both may feature Fibrocystic changes at HRCT at presentation.

[9] And represent a considerable proportion of interstitial lung disease.

Material and Methods

In this retrospective study, performed between January and August 2012, a population of 162 patients with biopsy documented lung disease (Imaging and Research Unit of University of Athens Medical School, Evgenidion Hospital) were classified in two groups. Group A included 81 patients with pulmonaryPathology (51 women, 30 men, mean age 57.8 ± 12.3 years) and group B included 81 patients with Constant interstitial pneumonia -UIP- (23 women, 58 men mean age 69.3 ± 9.8 years) respectively. Patient selection included consecutive patients presenting with pulmonary fibrosis in which lung biopsy documented eitherPathology or UIP. High-resolution computed tomography scanning of the lung was evaluated; the images were obtained by using 1mm collimation every 10mm throughout the thorax. The scans were obtained having the patient in supine position at full inspiration and were reconstructed by using a high spatial-frequency algorithm. All images were viewed at window settings optimized for assessment of lung parenchyma. HRCT features were evaluated per lung lobe (ground glass, reticular pattern, micro nodular, thickened septal lines, ground glass with traction bronchiectasis and bronchiectasis, honeycomb, Hilary and mediastina lymph node enlargement). Quantification of the severity and extent of each feature was also performed and compared. Gay and Watters technique was used and all HRCT findings scored on a scale 0-5 [10]. These scores were also summed into a total CT score for each patient and for each disease separately. Hence the severity and distribution of HRCT features was

recorded regarding the 81 patients with Pathology and the 81 patients with UIP. Statistical analysis: Fisher's exact test was used to evaluate differences between the proportions. A value of $p < 0,05$ was considered statistically significant. Ethical considerations: This retrospective study complies with the Declaration of Helsinki.

Results

Of the 81 patients with Pathology, 52 (64.20%) presented a reticular pattern, 62 presented thickened septal lines (76.54%), 24 presented honeycomb pattern (29.63%), 52 ground glass attenuation (64.20%), 7 ground glass presented traction bronchiectasis and bronchiectasis (8.64%), 47 micro nodular pattern (59.26%) and 48 Hilary and mediastina lymph node enlargement (60.49%). The distribution and severity findings were summarized in Table 1; the reticular pattern had a peripheral distribution in 13 patients (25.00%), the central per bronchial distribution in 40 patients (76.92%) and the involved upper lobes in 38 patients (73.08%), the middle lobe in 29 patients (55.77%) and the lower lobe in 41 patients (78.85%). The severity score of parenchymal reticular involvement was 30.22%. 62 patients (76.54%) recorded thickened septal lines and 52 (83.87%) with peripheral distribution, 28 (45.16%) with central per bronchial distribution. 35 (56.45%) showed distribution in the upper lobes, 34 (54.84%) in the middle lobe and 43 (69.35%) in the lower lobe. The severity score was 23.06%. 52 patients (64.20%) recorded ground glass opacities and 33 (63.46%) with peripheral distribution, 14 (26.92%) with central per bronchial distribution. 20 (38.46%) showed distribution in the upper lobes, 36 (69.23%) in the middle lobe and 26 (50%) in the lower lobe. The severity score was 17.04%. Ground glass opacities associated with traction bronchiectasis and bronchiectasis were observed in 7 patients (8.64%). A peripheral distribution was recorded in 2 (28.57%), central per bronchial distribution in 3 (42.86%), involved upper lobes in 1 (14.29%), middle lobe in 4 (57.14%) and lower lobe in 6 (85.71%). The severity score was 2.47%. Micro nodular opacities were found in 47 patients (59.26%). A peripheral distribution was noticed in 36 (76.60%), central per bronchial distribution in 15 (31.91%), upper lobes involvement in 24 (51.06%), middle lobe in 33 (70.21%) and lower lobe in 29 (61.70%). The severity score was 15.65%. Honeycomb pattern was found in 24 patients (29.63%). The honeycomb pattern had a peripheral distribution in 24 (100%), central per bronchial distribution in 5 (20.83%) and upper lobes involvement in 14 (58.33%), middle lobe in 16 (66.67%) and lower lobe in 21 (87.50%). The severity score was 14.02%. Mediastina and Hilary lymph node enlargement was found in 48 patients (60.49%). Overall, the severity of parenchymal involvement was 26 ± 19 . Figure 1 shows the distribution of the severity or parenchymal involvement in the patients of Pathology and it clearly reveals that the severity is mild in these patients (Figure 1). The semi quantitative severity assessment showed mild involvement (score 0-49) in 73 patients (90.12%), and mild severity (score 50-99) in 8 patients (9.88%) respectively.

Discussion End-stage pulmonary Pathology is typically characterized by severe interstitial fibrosis. The Fibrocystic stage of Pathology and idiopathic interstitial fibrosis concerns the 60% of fibrosis [11]. In this study, we examine the differences between pulmonary Pathology and UIP with high-resolution computed tomography (HRCT) regarding imaging features and quantification of the severity and extent of the disease. The study compared the HRCT findings per lobe separately. According to our study, micro nodules

were 59.26% in Pathology and 30.86% in UIP ($p=0.0335$) respectively. We found that the distribution in Pathology was greater in middle zone with the micro nodules located bilaterally. Nuns H et al refer that nodules are the hallmark of pulmonary Pathology, seen in 80% to 100% of all patients at HRCT but less frequently in stage IV [12]. In our study, nodules were small, and had a pattern of irregular and poorly circumscribed margins. A per lymphatic distribution of micro nodular lesions is the most common parenchymal disease pattern seen in patients with pulmonary Pathology. Per bronchial distribution was seen in 31.91% with an upper lobe predominance in 51.06%, showing a slight differentiation from the commonly described patterns. A fact that may indicate, that when Pathology is represented at first diagnosis with parenchymal involvement, the distribution is less typical. HRCT literature shows small nodules (2-4 mm in diameter), Constantly with a bilateral and symmetrical distribution, predominantly but not invariably in the upper and middle zones [13]. The severity score of micro nodules was low in our study, reaching a 15.65%. A percentage, that is in concordance with the literature [3]. The micro nodules were found most often in the sub pleural per Broncho vascular interstitial and less often in the septal lines. Hunninghake et al [14], in their study concluded that the lower lung honeycombing and the upper lung irregular lines were the only independent predictors and, by using only these two factors, a diagnosis of UIP could be established with a sensitivity of 74%, a specificity of 81%, and a positive predictive value of 85%. Other studies have consistently found that honeycombing is the best discriminator between UIP and other conditions [15]. We found honeycomb pattern in 69.14% of patients with UIP and 29.63% in patients with pulmonary Pathology ($p=0.0037$). The distribution was mostly peripheral. Abe sera et al showed that the Fibrocystic changes of pulmonary Pathology, particularly bronchial distortion and honeycombing, were in the upper and middle zones in most patients. In our study, patients with Fibrocystic Pathology were biopsy-documented with non-caveating granulomas and absence of fibroblastic foci characteristic of Pathology and UIP respectively, excluding therefore the possibility of coexistence of these entities. In addition, on follow up of the patients the diagnosis did not change based on clinical observations or treatment response. Bronchial distortion was also mainly central, and honeycombing was peripheral. We observed fibrosis in upper, middle and lower lobes. Honeycomb on CT is an important prognostic determinant in patients with UIP. Got way et al [16] declare that Flaherty et al [17,18] refer to a definite (confident) HRCT diagnosis of UIP on the basis of basilar honeycombing which portends a worse survival for individuals without honeycombing on the HRCT than for those with a histopathological diagnosis of UIP. In our study, ground glass opacities associated with traction bronchiectasis and bronchiolectasis presented a statistically significant correlation to UIP ($p = 0.0021$) and the distribution was mostly central. In the study by Baughman et al [19] traction bronchiectasis tend to be an upper lobe predominant process and can have relative sparing of the lower lobes in pulmonary Pathology, whereas we recorded a lower lobe predominant process. When ground glass opacities are associated with reticular lines or traction bronchiectasis, they Constantly indicate histologic fibrosis [20]. Basher et al study showed that thickened septal lines were obviously Fibrocystic due to their irregularities, angulations and association to some signs of fissure and bronchial distortion [21]. Previous studies have shown that they were irreversible [22]. In our study, thickened septal lines seem to be more frequent to UIP compared to Pathology ($p = 0.05$). Fibrocystic lesions in UIP are often located in the periphery of the secondary pulmonary lobules, adjacent to septal lines. Lynch A D et al refer that scans considered

consistent with IPF, were significantly more likely to show honeycombing, traction bronchiectasis and bronchiectasis, and lower lobe volume loss, with less likelihood to show ground glass attenuation, decreased attenuation, mosaic attenuation, and centrilobular nodules compared to scans which were not consistent with IPF [23]. We found 60.49% enlargement of lymph nodes in Pathology and 1.23% in UIP at a statistically significant level ($p = 0.0001$). In Pathology, the enlargement lymph nodes were Hilary and mediastina, bilateral and symmetric. This is the most common pattern in Pathology. Baughman et al supported that adenopathy can help in distinguishing Pathology from other interstitial lung diseases [19]. The presence of adenopathy alone or along with parenchymal disease has been found in more than half of Pathology patients at the time of diagnosis. CT scan is far more sensitive for detecting adenopathy. Various interstitial lung diseases can cause some adenopathy, but the enlargement is Constantly present only to a mild extent [19]. Gay and Watters' HRCT scoring system uses a scale of 0-5 for ground glass opacity and linear opacity and estimates the severity and the extent of each disease [10]. In our study, we estimated all HRCT findings, per each lobe separately and assessed the severity and extent of pulmonary Pathology and UIP and then we calculated the total severity for each patient. UIP in an adequate population of patients presents moderate severity of disease, while in pulmonary Pathology the majority of patient's exhibit mild severity. Shigemitsu et al [11] observed that when patients with Pathology showed rapid progression of the disease, they were ultimately led to lung transplantation. Our work revealed that differences in the severity of involvement between the two disease entities and indicated that more severe degree of disease is present in UIP. The severity of each disease is related to the patient's prognosis. Study limitations include the retrospective nature of the study, and the relatively small number of patients. In conclusion, the major discriminators between the two entities at presentation is the micro nodular pattern favoring Pathology, the honeycomb pattern indicating UIP, the ground glass opacities with traction bronchiectasis favoring UIP and the lymph node enlargement which most frequently found in Pathology. Severity scores are more advanced in UIP.

Financial Disclosure and Conflicts of Interest All authors declare that there is no conflict of interest.

References

- [1] Rockoff S D, Rohatgi K. Prashant. UnConstant manifestations of thoracic Pathology. A J R 144: 513-528.
- [2] Lynch JP 3rd, White ES. Pulmonary Pathology. Eur Respir Monograph 32: 105-129.
- [3] Criado E, Sanchez M., Ramirez J, Arguis P, de Caralt MT, Perea JR, Xaubet A. PulmonaryPathology: Typical and atypical manifestations at High-Resolution CT with pathologic correlation. RadioGraphics 30: 1567-1586.
- [4] Baughman RP, Winget DB, Bowen EH, Lower EE. Predicting respiratory failure inPathology patients.Pathology Vasc Diffuse Lung Dis .
- [5] Hennebicque AS, Nunes H, Brillet PY, Moulahi H, Valeyre D, Brauner MW. CT findings in severe thoracicPathology. Eur Radiol ; 15: 23-30.