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Prevalence of Diffuse Parenchymal Lung Disease (DPLD) and Associated Fibrosis in Northern Saudi Arabia

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Abstract: ***Background: Diffuse parenchymal lung diseases (DPLD) is a wide spread disorders including occupational or environmental exposure, therefore, the objective of this study was to estimate the prevalence of DPLD in Northern KSA. Methodology: In this study clinical, imaging and laboratory parameters were obtained from 60 patients diagnosed with DPLD in the Pulmonary Medicine Department at King Khalid Hospital. Results: Of the 60 patients, DPLD was Identified in 17/60 (28.3%) patients of whom 16/17 (94%) were idiopathic pulmonary fibrosis (IPF) with and 1/17 (6%) with extrinsic allergic alveolitis (EAA). Conclusion: DPLP is prevalent in Northern KSA. Further investigation in regard to etiology are urgently required.***

Keywords: Diffuse parenchymal lung disease, Saudi Arabia, idiopathic pulmonary fibrosis

1. Introduction

Diffuseparenchymallung diseases (DPLD) constitutes a miscellaneous group of disorders affecting the distal lung parenchyma, exactly the tissue and spaces surrounding the alveoli. These tissues may be filled with inflammatory cells, proliferating fibroblasts or established fibrosis, which leads to architectural distortion and impaired gas exchange. Although the underlying pathogenetic mechanisms are known or inferred for some DPLD (such as sarcoidosis, silicosis, drug reactions and collagen vascular diseases), the pathogenesis of the majority of these entities, particularly those categorized by progressive fibrosis, is poorly understood[1].

DPLD are believed to be *complex* diseases, causedby genetic variations involving multiple genes, each contributing an effect of variableextent. However, a person may have the needed genetic profile to develop a disease and yet it will not noticeable unless an environmental or infectious factor is met[2]. DPLD covers a varied range of diseases, and now the diagnosis is based on a combination of clinical, physiologic, radiologic, and, if required, histopathologic criteria. In spite of developments in medical therapy and technology, the prognosis in Idiopathic Pulmonary Fibrosis (IPF) remains obstinately poor, and the need for progress and new methodologies is important. The deficiency of progress reflects the poorly understood etiology and pathogenesis of DPLD and IPF[3-6].IPF is the most common type of the idiopathic interstitial pneumonias. The histopathologic hallmark is a heterogeneous appearance in which areas of fibrosis with scarring and honeycomb change alternate with areas of less affected or normal parenchyma[7]. Since the prevalence of DPLD differs from community to another and from country to another, the objective of this study was to estimate the prevalence of DPLD in Northern KSA.

2. Materials and Methods

In this study, study subjects were identified by a retrospective retrieval of all patients who had undergone clinical, laboratory and radiological investigations atking Khalid Hospital for the diagnosis of pulmonary fibrosis from January 2014toJanuary 2015. All patients had suspected with interstitial lung disease based on clinical information, laboratory and radiological testing were included. Clinical parameters, radiography and biopsy results together with demographical data were identified and analyzed using SPSS software (version 16) to identify frequencies and to cross tabulate different parameters.

The fibrosis score was analyzed using the hematoxylin-and-eosin-stained lung section slides. The extent of fibrotic lesions was scored as 0 (0-10%), 1 (10-25%), 2 (25-50%), 3 (50-75%), or 4 (75-100%). The severity of fibrosis was scored from 0 (normal lung) to 8 (total fibrosis) by determining the average of 10 distinct microscopic fields at 200× magnification in accordance with the Ashcroft scoring system[8].

3. Results

In this study 60 patients were identified as having pulmonary lung fibrosis during the period from January 2010 to January 2014, their ages ranging from 15 to 96 with a mean age of 58 years. Of the 60 patients, 33/60(55%) were males and 27/60(45%) were females, giving males' females' ration of 1.22: 1.00. Of the 60 patients with lung fibrosis, 17/60 (28.3%) were identified with DPLD. Of the 17 patients with DPLD, 16/17 (94%) were found with IPD and the remaining 1/17 (6%) was identified with extrinsic allergic alveolitis. Out of the 17 patients, 11/17 (64.7%) were males and 6/17(35.3%) were females. The risk associated with male sex with 95% confidence interval giving an odd ratio (OR) of 1.75(0.5483 to 5.5859). As

indicated in Table 1, the majority of cases of DPLD were identified at age group 56+ constituting 14/17 (82.3%). Consequently, the risk of DPLD is significantly increase with the increase of age (P <0.0001).The mean age at diagnosis was 55 years. In regard to the age sex, although, relatively most cases were seen at elder age for both sex, but some younger males were found as indicated in Fig.1.

Table 1: Distribution of the study population by DPLD and age

Age group	PDLD		Non-DPLD	Total
	IPD	EAA		
<25 years	1	0	6	7
26-35	1	0	4	5
36-45	1	0	4	5
46-55	0	0	5	5
56+	13	1	24	38
Total	16	1	43	60

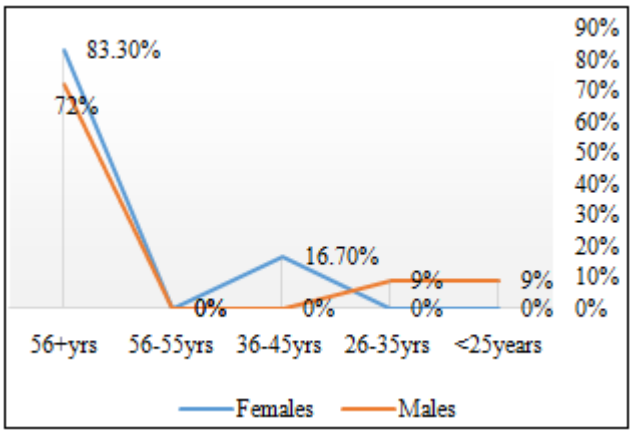

Figure 1: Description of the study population by age and sex

However, some of the patients with DPLD were found with other conditions including; Arrhythmia (11.8%); Cardiovascular CVS (35.3%), Diabetes Mellitus (DM) (64.7%), Hepatic (5.9%), Obesity (11.8%), Malignancy (5.9%) and Death occurred in 5.9%, as shown in Fig 2.

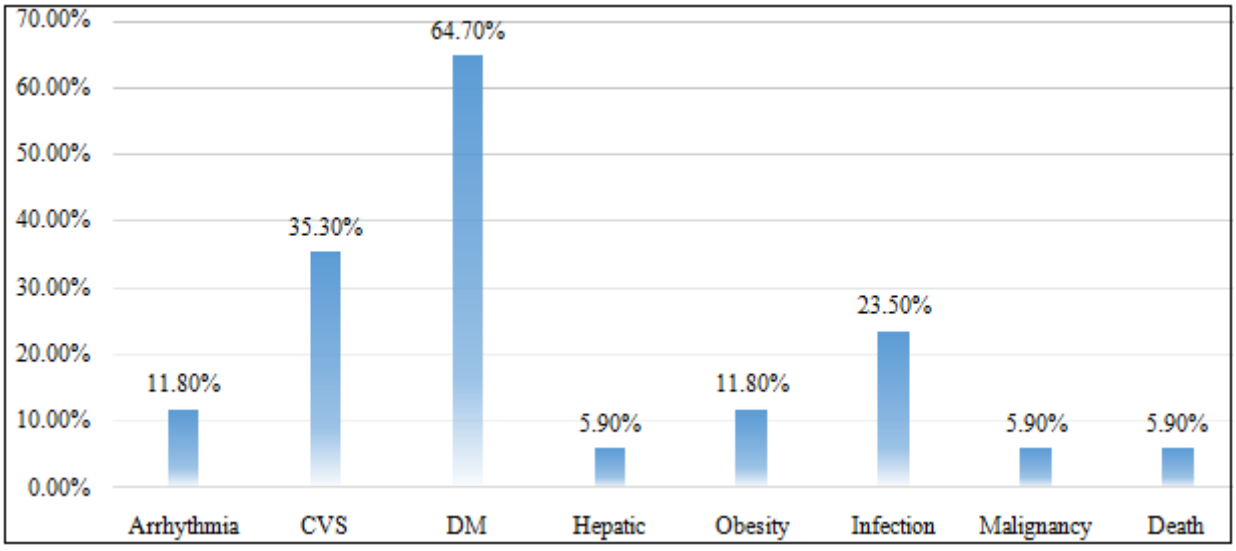

Figure 2: Description of the DPLD by other associated conditions

In regard to the scording of the severity of lung fibrosis; 5 cases identified in score 0, 3 cases score 2, 5 cases score 3, 2 cases sore 4, as indicated in Fig 3.

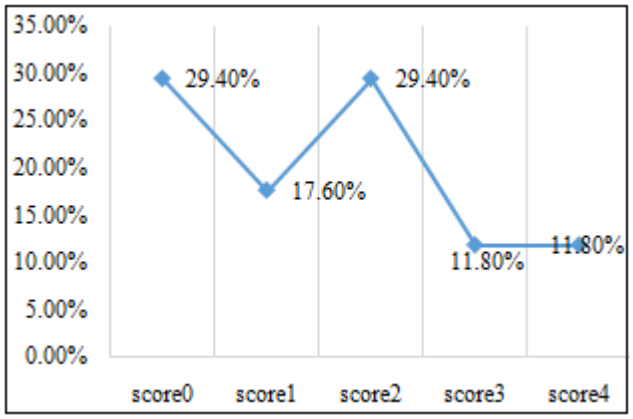

Figure 3: Description of the DPLD by score of the severity of lung fibrosis

4. Discussion

DPLD is a fatal lung disease of in most instances of unknown causes, particularly idiopathic pulmonary fibrosis type. However, in this study, the burden of DPLD was assessed in a series of patients referred to the department of pulmonary medicine with lung disorders. In the present study, the prevalence of DPLD was found to 28.3%, in thin the other causes of lung fibrosis which is relatively high. The interstitial lung diseases are a heterogeneous group of disorders characterized by inflammation and/or fibrosis of the pulmonary interstitium. In 2002, the American Thoracic Society and the European Respiratory Society revised the classification of interstitial lung diseases and introduced the term diffuseparenchymallung disease (DPLD). The idiopathic pulmonary fibrosis type is a subtype of diffuseparenchymallung disease[9].

Nevertheless, in the present study 94% of the cases of DPLD were cases of IPF. IPF is the most common idiopathic

interstitial pneumonias with an estimated 5-year survival of approximately 20%. The disease seems to occur following chronic injury by abnormal/dysfunctional alveolar epithelial cells that stimulate fibroblast recruitment and proliferation, resulting in scarring of the lung and permanent loss of function[10].Data on IPF are limited yet show extensive differences in different worldwide[11].There is a lack of Data from KSA, regarding IPF, and most of those studies dealt with treatment choices[12]. A study from Saudi Arabia included 330 patients with interstitial lung disease (ILD), out of which 23% had IPF[13]. Worldwide, the prevalence estimates for IPF range from 6 to 32 per 100,000[14].

In the present study, males were more affected with IPF than females and the disease was frequently seen among elder patients both in males and females. It was found that, the males with IPF were more likely to be in their 60s[15]. Another study has shown that, In Saudi Arabia, IPF patients tended to be slightly older and the disease progression was somewhat slower than reported IPF cohorts in other populations[14]. Moreover, increased number of females' patients over that of males was previously reported[16]. Studies have found that, IPF patients in Saudi Arabia are most often elderly, obese, and female [14]. Regard to the other conditions, DM which, common in among patients in this study, is supposed to increase the risk of IPF[17,18].

On the other hand, and according to the severity of lung fibrosis, there was increased percentage in score 2, though, there was relatively similar scoring in scores from 0 to 4. The limitation of this study wasits retrospective design, which prevent many associated factors to be exposed. In conclusion, DPLP is prevalent in Northern KSA. Further investigation in regard to etiology are urgently required. Genetic factors possibly play a role in IPF risk and progression. Geographic unevenness of IPF prevalence may be influenced by alterations in mortality, probably as a result of the influence of comorbidities such as diabetes, pulmonary hypertension, and cardiovascular disease.

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