IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 1 Ver. III (January. 2017), PP 15-19
www.iosrjournals.org

Combined Pulmonary Fibrosis and Emphysema - A Case Series

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Abstract
Background: *Idiopathic interstitial fibrosis and pulmonary emphysema are commonly associated, showing bilateral upper zone emphysema and bilateral fibrosis of lower lung fields. It's a progressive disease characterized by restrictive lung function and decrease gaseous exchange.*
Objective: *To differentiate this disease entity early from other fibrotic diseases. To start the treatment at early stage.*
Method: *A Case series of 9 patients diagnosed with CPFE disease at SBKS MIRC included in the study.*
Results: *Out of total 9 patients 8 were males and 1 female. Mean age was found to be between 40 to 76 years. Disease found to be more prevalent in smokers. Mean Spo2 on examination was found to be 88%.*
Conclusion: *CPFE is a distinctive syndrome which should be diagnosed at early stage preventing morbidity and early mortality along with providing better quality of life to patients . There is considerable prevalence of CPFE in smokers with increasing age too, which is always misdiagnosed, can lead to in fact worsening of patient`s condition.*
Keywords: *CPFE, misdiagnosis, adult-onset, senile, elderly, copd.*

I. Introduction

Combined Pulmonary Fibrosis and Emphysema (CPFE) is a syndrome characterised by Upper lobe emphysema and bilateral Lower lobe fibrosis. This coexistence of pulmonary fibrosis and emphysema was first described in 1990 by Wiggins et al.[1] The term CPFE was coined by Cottin et al in 2005 as a syndrome of heterogeneous population of patients which is diagnosed by high resolution computed tomography (HRCT). This disease is rare. Emphysema and pulmonary fibrosis have dissimilar physiologic effects. Emphysema causes reduced lung elastic recoil, increased lung compliance, and increased lung volumes with reduced maximal expiratory flow rates, whereas pulmonary fibrosis results in increased lung elastic recoil, decreased lung compliance, and reduced lung volumes with preserved or even increased maximal expiratory flow rates at a given lung volume. Clinically, either emphysema or fibrosis typically predominates, and individual patients are rarely recognized as having both disorders simultaneously[2]. Combined pulmonary fibrosis and emphysema is common in smokers. High-resolution CT (HRCT) scanning has enhanced clinical recognition of the simultaneous occurrence of emphysema and pulmonary fibrosis. Radiographic characteristics include the presence of upper-lobe emphysema and lower-lobe pulmonary fibrosis.

A single gene defect responsible for most cases of CPFE has not been established in humans.[2] in non smokers certain mutations in surfactant related genes like ABCA3 which has a role in surfactant storage and homeostasis has been identified.[3]

There has been few case reports since then and an even more few case series and even more so from India.

II. Materials & Methods

- Crossectional study.
- Total of 9 patients were included in study, visiting Department of Respiratory Medicine, SBKS MI & RC, Vadodara on outpatient and inpatient basis were included.
- All included patients had undergone chest x ray, spirometry, HRCT.

Inclusion criteria:

- Patients with clinical symptoms and confirmed diagnosis by HRCT showing parenchymal lesion with fibrosis, bilateral bibasilar reticular opacities with honeycombing/ground glass opacities tractional bronchiectasis were included.

Exclusion criteria:

- Patients with other connective tissue disorders.
- Patients with other known causes of ILD were excluded.

At the end of this study, the collected data was analyzed by the appropriate statistical methods whenever required such as Percentage, Mean, Median, Mode, Standard deviation, Confidence interval, Correlation, Sensitivity etc.

III. Results

In our case series a total of nine cases were included. Of the nine cases majority of the cases were males and total percentage was 89%(8) (Figure 1). The mean age of the patients in our study was 60 years ranging from 40 years to 76 years (Figure 2).

In our study nearly 88% of the patients were found to be smokers (male + female) (Figure 3).

45 % of the patients were Farmers followed by Drivers and labourers of 22% each followed by 11% of house wife. All patients presented with complaints of cough and breathlessness. On examination 78% of patients presented with clubbing and Mean SPO_2 was 88% (Figure 4). All patients on auscultation were found to have bibasilar Velcro type crepitations associated with wheeze.

All patients had undergone respective x rays and HRCT. On chest x rays we got hyperinflation in bilateral upper zones and reticular nodular opacities in bilateral lower zones (Figure 5-A). This finding was further confirmed by HRCT in which we got found panacinar and paraseptal emphysema noted in bilateral upper lobes, honeycombing , ground glass opacities and tractional bronchiectasis in lower lobes (Figures 5-B & 5-C).

IV. Discussion

Combined pulmonary fibrosis and emphysema is a rare syndrome and this term has been in use since Wiggins described it in 1990.

Our study is a case series of 9 patients who were selected on basis of HRCT which showed panacinar emphysema noted in bilateral upper lobes left upper lobe , interstitial fibrosis seen in bibasilar regions, Of these patients there was a male predominance of 89% and of the mean age group of 60 years.

Many studies already been indicated that patient exhibiting connective tissue disorders are younger in age with female predominance and they also have less DLCo impairment as compare to the patients encountered with idiopathic CPFE cases.[4]

As per the systematic review by Matthew D. Jankowich and Sharon I. S. Rounds CPFE is more common in Smokers (98%) and has a male sex predominance (90%) , which is similar to our study where we have got 89% of the patients as smokers.[2]

Study by Washko GR et al. also suggested that smokers have high incidence of restrictive pattern on spirometer and HRCT attenuation which is similar to our study also there is male sex predominance of 8 out of 9 being male patients and 8 out of 9 being smokers and the female was a smoker with restrictive pattern on spirometer.[5]

In a case series of 61 patients done as a multicentre study in France the mean age was 65.2 years. In our study the mean age was 60 years. And in another study of 10 patients by Alilovic M et al had their patient mean age of 68 years which is nearly similar to the study age group of our patients. In their study the mean spo2 of patients were 85% where as in our study it was 88% ranging from 82% to 95%.[6]

In a systematic review the survival period of the disease has been discussed and it has a median ranging from 2.1 to 8.5 years. And if pulmonary hypertension has been confirmed then the survival period of 1 year is only 60%.[2]

In our study we haven't followed up the patients for their mortality hence it should be done and need to be carried out for mortality and morbidity.

V. Conclusion

Why it is of importance to diagnose the CPFE syndrome?

- ✓ Early demarcation of disease from other entities should be done at the earliest by specialists, as HRCT exhibits clear distinction between emphysema and fibrosis and thick wall cystic lesions, by other means of suitable and relevant investigations.

- ✓ To diagnose disease at early stage and prevent development of pulmonary artery hypertension, as the prevalence of pulmonary artery hypertension seen to higher i.e. around 47% to 90%in these patients, thereby reducing the co morbid condition and improving quality of life for the patient.

VI. Declaration Of Interest / Conflict Of Interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References

[1]. Wiggins J , Strickland B , Turner-Warwick M . Combined cryptogenic fibrosing alveolitis and emphysema: the value of high resolution computed tomography in assessment . *Respir Med* . 1990 ; 84 (5): 365 – 369.

[2]. Jankowich M, Rounds S. Combined Pulmonary Fibrosis and Emphysema Syndrome. Chest. 2012;141(1):222-231.\

[3]. Epaud R, Delestrain C, Louha M, Simon S, Fanen P, Tazi A. Combined pulmonary fibrosis and emphysema syndrome associated with ABCA3 mutations. European Respiratory Journal. 2013;43(2):638-641.

[4]. Cottin V, Nunes H, Mouthon L, Gamondes D, Lazor R, Hachulla E et al. Combined pulmonary fibrosis and emphysema syndrome in connective tissue disease. Arthritis & Rheumatism. 2010;63(1):295-304.

[5]. Washko G, Hunninghake G, Fernandez I, Nishino M, Okajima Y, Yamashiro T et al. Lung Volumes and Emphysema in Smokers with Interstitial Lung Abnormalities. New England Journal of Medicine. 2011;364(10):897-906.

[6]. Alilović M, Peroš-golubićić T, Tekavec trkanjec J, Hećimović A, Smojver-ježek S. Combined pulmonary fibrosis and emphysema syndrome. Lijec Vjesn. 2015;137(1-2):22-6.

Figures

Figure 1: Sex Distribution CPFE patients

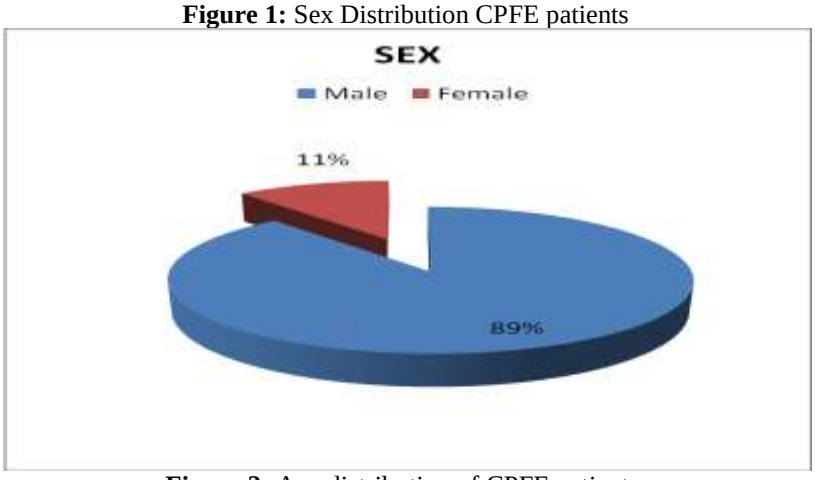


Figure 2: Age distribution of CPFE patients.

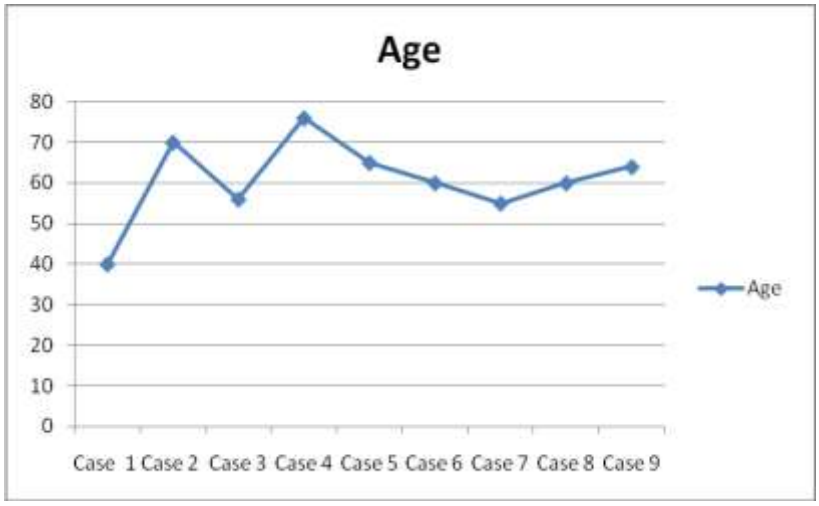


Figure 3: Smoking and CPFE

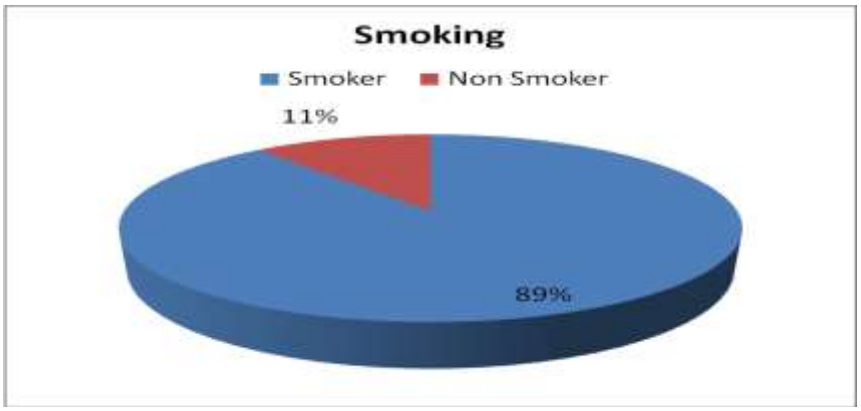


Figure 4: SPO_2 and CPFE

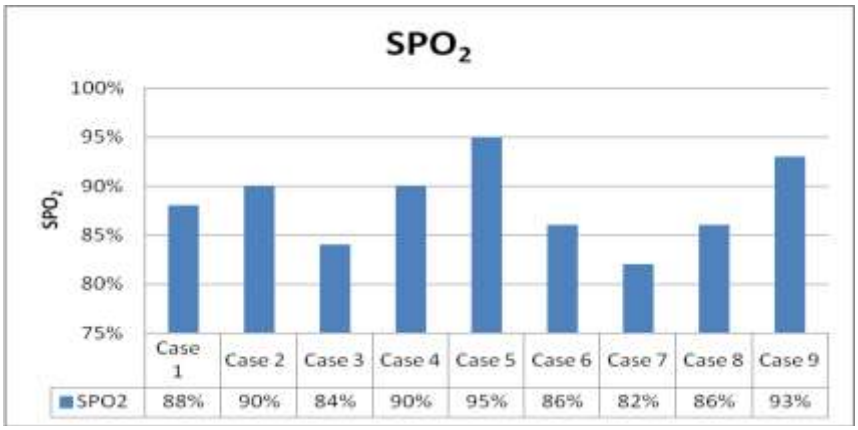


Figure 5-A Chest X Ray:

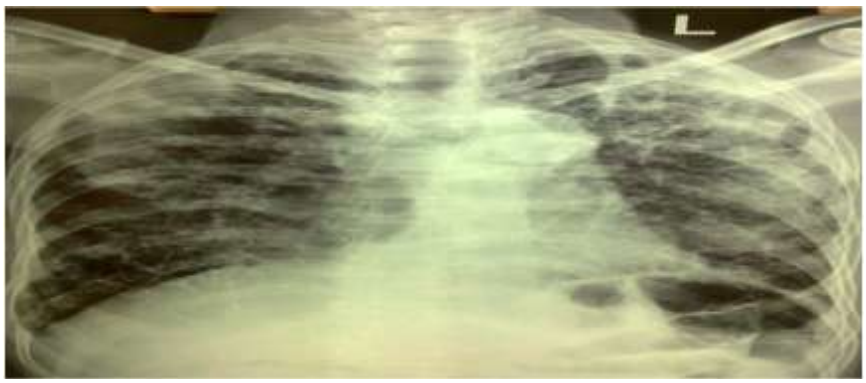


Figure 5-B Hrct – Bilateral upper lobe bullae:

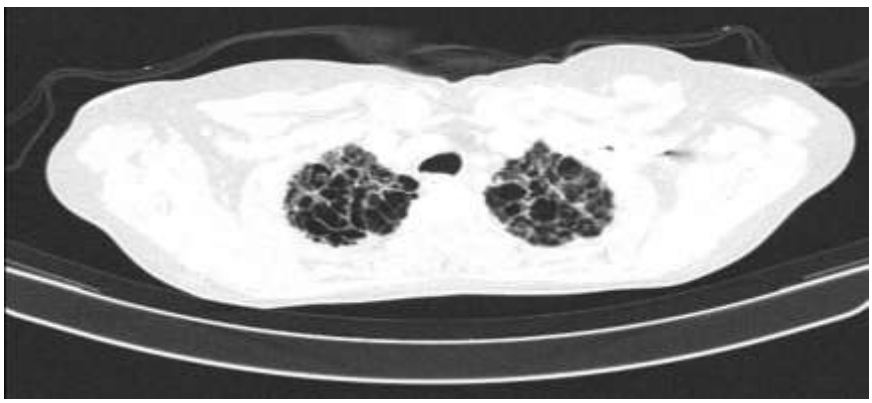
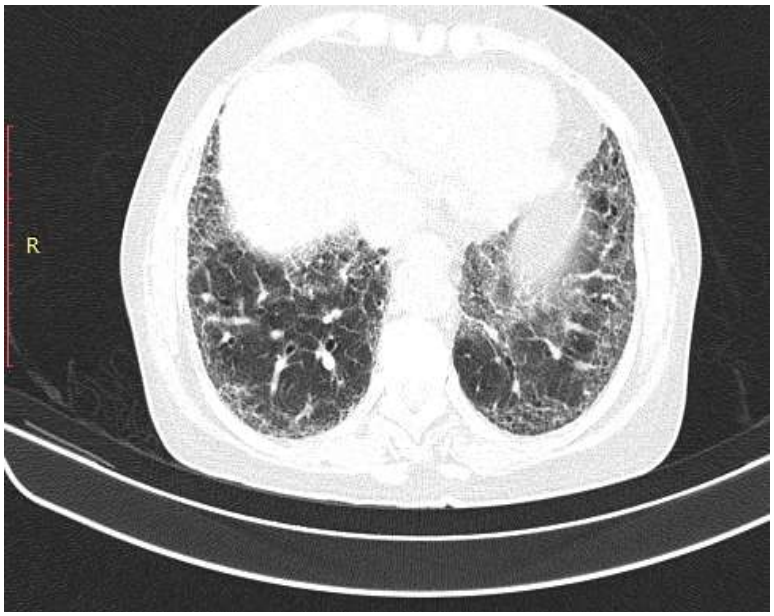


Figure 5-C Hrct Bilateral lower lobe fibrosis: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