ISSN No:-2456-2165

A Case Series on Combined Pulmonary Fibrosis and Emphysema

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Abstract:- In 2005, Cottin *et al.*, put forth a term, *Combined Pulmonary Fibrosis and Emphysema* (*CPFE*) which is a rare respiratory disorder and is characterized by exertional dyspnoea, upper-lobe emphysema and lower-lobe fibrosis, preserved lung volume and severely diminished capacity of gas exchange.[1][2] The main etiologies behind CPFE are heavy smoking history, hypoxemia, unexpected subnormal lung volumes and severe reduction of carbon monoxide transfer. High-resolution CT (HRCT) is the mainstream diagnostic parameter for CPFE. Apart from HRCT, spirometry values are also used to assess the severity of the disease.[4] Treatment options include symptomatic therapy as there is no specific treatment available till date, and also includes smoking cessation and oxygen therapy.[3] This case series involves 3 cases of CPFE with different symptoms and treatment has been given accordingly.[8]

Keywords:- *Combined Pulmonary Fibrosis and Emphysema (CPFE), Hypoxemia, HRCT, Spirometry.*

I. INTRODUCTION

Combined pulmonary fibrosis and emphysema (CPFE) is a rare pulmonary condition characterized by the involvement of both upper lobe emphysema and lower lobe fibrosis with very low diffusion capacity in contrast with subnormal spirometry that occurs mainly in heavy smokers with severe dyspnoea and exercise limitation.[1][6] Cough and dyspnoea are common symptoms in patients with CPFE or Chronic Obstructive Pulmonary Disease (COPD) or Idiopathic Pulmonary Fibrosis (IPF).[7] From several studies it has been found that a person with an already existing COPD when exposed to cigarette smoke becomes vulnerable to developing emphysema and pulmonary fibrosis. The high-resolution computer tomography (HRCT) scanning has been adopted as the main diagnostic method for CPFE. The HRCT would typically show centrilobular or paraseptal emphysema which is often predominant in the upper zone.[4] The desired treatment option for a CPFE patient is a long term oxygen therapy. Anti-fibrotic drugs (Pirfenidone, Nintedanib) have proven to relieve CPFE symptoms to a limit.[7]

❖ *CASE 1*

A 74-year-old female patient was admitted in the respiratory department of a tertiary care hospital. The patient had complaints of cough with sputum, wheezing, chest congestion for 1 month. The patient's history showed that she was exposed to biomass fuel and had dust allergy for several years. She was a known case of Type 2 Diabetes Mellitus (DM), Hypertension (HTN), Sinusitis, Coronary Artery Disease (CAD) and Obstructive Sleep Apnoea (OSA). She had a history of Total Knee Replacement (TKR) surgery in 2014.

On examination the patient's vitals were as follows:

PARAMETERS	VALUES
BP	140/90 mmHg
BODY WEIGHT	68 kg
RESPIRATORY RATE	20 breaths/min.
PULSE RATE	65 beats/min.
SPO_2	97%

Table 1

➢ *Chest HRCT Examination:*

- Heterogenous lung attenuation with areas of air trapping and mosaic perfusion – likely secondary to obstructive lung disease.
- Multiple peripherally placed paraseptal bullae in the upper lobe.
- There is evidence of multiple contiguous rows of peripherally located lung cysts in the basal distribution, showing adjacent mild fibrotic component and subtle traction bronchiectasis – *Findings likely to represent honey combing.*

Therapeutic management of this condition includes rest, periodic assessment of oxygen saturation (SPO_2) and oxygen supplementation. Patient was initially treated with Foracort 200 MDI (Budesonide 200 mcg + Formoterol 6 mcg) at a dose of 1 puff/twice daily which has to be used with Zerostat VT spacer. After administering this, the patient developed slight tremor and palpitation which resolved eventually on its own. The patient was prescribed with T. ABFLO (Acebrophylline) 100mg, T. Montek LC (Levocetirizine 5mg

+ Montelukast 10mg), T. Medrol (Methyl Prenisolone) 8mg and Syp. Lupituss (Levocloperastine) 10ml for symptom relief.

❖ *CASE 2*

A 83-year-old male patient was admitted in the respiratory department of a tertiary care hospital. The patient had complaints of increased shortness of breath for 3 days, wheezing and cough with mucoid sputum for 3 weeks. He had a history of hemoptysis 6 months back and chest discomfort was observed. The patients had comorbidities like CAD, Non ST-elevated Myocardial Infarction (NSTEMI), severe aortic stenosis, anemia, old Pulmonary Tuberculosis (PTB), COPD. The patient was an ex-smoker as well as an ex-alcoholic.

On examination the patient's vitals were as follows:

PARAMETERS	VALUES
BP	120/80 mmHg
RESPIRATORY RATE	22 breaths/min.
PULSE RATE	98 beats/min.
SPO_2	96%

Table 2

➢ *Chest X-ray:*

Chest x-ray showed features of bilaterally scattered fibrosis. Hence HRCT was taken.

➢ *Chest HRCT Examination:*

- Extensive centriolar emphysema bilaterally with predominant involvement of upper lobes.
- Mild fibrotic bands with traction bronchiectasis – *Findings likely to represent honey combing.*

Therapy for this condition includes rest, periodic assessment of oxygen saturation (SPO_2) and oxygen supplementation. Patient was initially treated with Neb. Foracort 0.5mg (Budesonide 500mcg + Formoterol 20mcg), Neb. Levolin 1.25mg (Levosalbutamol), Neb. Ipravent (Ipratropium Bromide). The patient was prescribed with T. Telekast F (Montelukast 10mg + Fexofenadine 120mg), T. Mucinac (Acetylcystiene), T. Doxovent (Doxophylline) for symptom relief. The patient later improved and thus discharged with following medications T. Sompraz (Esomeprazole) 40mg, Neb. Foracort 0.5mg, Neb. Duolin 2.5ml (Levosalbutamol, Ipratropium), T. Doxovent 400mg, T. Mucinac 600mg, T. Ivepred (Methyl prednisolone) 8mg, T. Telekast 40mg.

❖ *CASE 3*

A 82-year-old male patient was admitted in the respiratory department of a tertiary care hospital. The patient had complaints of fever, nausea, vomiting, cough with whitish expectoration since 2 days. He was a known case of Type 2 Diabetes Mellitus (DM), Hypertension (HTN), Dyslipidemia (DLP), CAD and COPD. The patient's lab report showed an abnormally high CRP.

On examination the patient's vitals were as follows:

PARAMETERS	VALUES
BP	140/90 mmHg
BODY WEIGHT	68 kg
RESP. RATE	20 breaths/min.
PULSE RATE	65 beats/min.
SPO_2	97%

Table 3

➢ *Chest X-ray:*

Chest x-ray showed infiltration in the right lungs.

➢ *Chest HRCT Examination:*

- Diffused intralobular septal thickening predominantly in bilateral lower lobes with multiple fibrotic bands showing secondary traction bronchiectasis and tiny cystic lucencies arranged in multiple row like configuration towards basal segments of bilateral lower lobes associated with minimal bilateral nodular pleural thickening.
- *Findings likely to represent idiopathic pulmonary fibrosis.*
- Tiny paraseptal bullae in the bilateral upper lobes and right middle lobe suggestive of background emphysema.

Therapeutic management of this condition includes rest, periodic assessment of oxygen saturation (SPO_2) and oxygen supplementation. Patient was initially treated with Neb. Duolin, Neb. Budecort (Budesonide) 0.25mg and Inj. Viatran (Cefoperazone 2g + Sulbactam 1mg) 3g and T. Azithral (Azithromycin) 500mg. The patient was prescribed with T. Mucolite for symptom relief.

II. DISCUSSION

CPFE is a rare pulmonary condition characterized by the involvement of both upper and lower lobe with emphysema and fibrosis respectively. Cough and dyspnoea are the most common symptoms that can be seen in patients with CPFE. This condition can be diagnosed and confirmed with the help of spirometry values and HRCT impressions. The HRCT would show honey combing structures as well as centrilobular or paraseptal emphysema in patients with CPFE. As there is no specific treatment available till date, the

condition can be managed with long term oxygen therapy along with other medications which will provide symptomatic relief.

This case series of CPFE showed three patients who were confirmed with the condition by help of HRCT impression. The treatment with corticosteroids and bronchodilators are effective in improving the clinical course of patients with CPFE. All the patients above showed a reduction in their oxygen saturation. Therefore, correction of this is the most important aim in the treatment of CPFE. Here the patients were advised to follow proper usage of Meter Dose Inhalers (MDI) and nebulizers for improved quality of life. The patients who were prescribed with nebulizers showed slightly more response than the patients who got inhalers in their therapy. Therefore, nebulizers have a more significantly curative effect, as it can effectively improve symptoms.

III. CONCLUSION

CPFE is a distinct pulmonary condition, so it is important to recognise the severity of various pulmonary symptoms associated with this condition. It will influence the patient's physical and social well being. Smoking may be the prime etiological factor for causing emphysema or fibrosis dominant. CPFE patients tend to exhibit a delay in the reduction of FVC and monitoring disease progression and therapeutic response to anti-fibrotic patients can be challenging. It is extremely important to identify and urgently refer potential severe cases in order to have the appropriate investigations and have the appropriate care administered.

IV. CONFLICT OF INTEREST

The authors declare no conflict of interest.

V. ABBREVIATIONS

CPFE	Combined Pulmonary Fibrosis & Emphysema
COPD	Chronic Obstructive Pulmonary Disease
IPF	Idiopathic Pulmonary Fibrosis
HRCT	High-Resolution Computer Tomography
DM	Diabetes Mellitus
HTN	Hypertension
CAD	Coronary Artery Disease
OSA	Obstructive Sleep Apnoea
TKR	Total Knee Replacement
DLP	Dyslipidemia
BP	Blood Pressure
SPO_2	peripheral capillary oxygen saturation
MDI	Meter Dose Inhaler

Table 4

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