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A 43 year old Woman with Shortness of Breath and Rare Interstitial Lung Disease - A Case Study

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ABSTRACT

In this study, interstitial lung disease (ILD), the term used to refer certain diseases that cause damage (scarring) of lungs. The scarring caused by ILD is often irreversible and it can be classified based on different criteria (known cause, unknown cause, acute chronic, granulomatous, non-granulomatous, history of habitual activities like smoking, no smoking etc.). There are other things that cause or increase the risk of ILDs includes certain drugs, genetics or certain type of medical treatments like radiation or chemotherapy. The ILD is very much linked with the exposure to asbestosis and hypersensitivity pneumonitis. In this, we present one of the rarest cases of ILD in a non-smoking young woman who had a progressive growth of ILD and passed away in 2019.

Keywords: Interstitial Lung Disease, CT scan, Lung Bronchoscopy

Introduction

People with autoimmune diseases are very much exposure to increase the risk of developing ILD's. In some cases, the causes are undefined and they are all referred as idiopathic pulmonary fibrosis. Some of the medications may slow down the damage of lung caused by ILD, but regaining of the full use of lungs are not possible at any stage. Lung transplantation may be an option for some people who have ILD based on their health condition and various parameters. ILDs can be classified broadly based on unknown and known cause, connective tissue disorders, drug induced interstitial lung disease. Fig. 1. Shows the different types of ILDs based on the above factors

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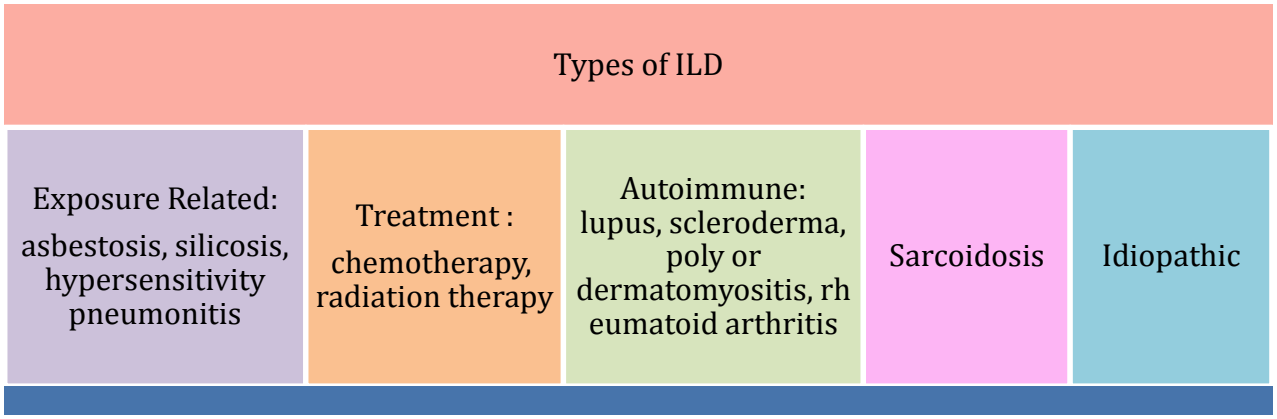

Fig. 1. Types of ILD

Interstitial Lung disease can also be described as DLD (Diffuse Lung Disease) that involves disorders in pulmonary parenchyma and interfere with the oxygen and carbon dioxide exchange. ILD includes extensive alteration of alveolar and the architecture of airway

Symptoms of ILD and the steps to be taken

Shortness of breath is the common symptom of all the ILD's and it is accompanied by chest pain, dry cough, tightness in the chest, fatigue and sometimes weight loss. Most of the cases, the symptoms appear only when the lung has severely affected and in some cases it can develop life threatening complications. Treatment of ILD include chest X-ray or CT scan initially and the total capacity of the lung may be measured using lung function test. From the Lung function test, the level of deformation of lungs may be measured and based on that next level of treatment may be started.

To diagnose a specific ILD and for serious cases, different invasive procedures such as lung biopsy or a bronchoscopy may be carried out. Based on the severity and the type of ILD, the treatment also varies from patient to patient. Since lung damage by ILDs are irreversible and also progressive in nature, so treatment may be on reducing the initials symptoms and to slowdown the progression thereby increasing the life time. Some sort of medication, oxygen therapy and pulmonary rehabilitation may also be recommended to improve the quality of life and to slow down the progression rate. Lung Transplantation may be suggested for those having extreme severity in the disease progression.

Case Report

A 43 year old married woman with a history of dry cough, shortness of breath and tightness in chest admitted in 2014 for initial diagnosis. Her family doctor suggested some normal antibiotics for the relief of the issue but after few days again she was presented with the same problem. This time, the general practitioner suggested her to consult a pulmonologist for further investigation. She was a non-alcoholic, non-smoker and no evidence for hazardous things.
There were no evidence of chest pain, rashes in the skin, pain in joints, tuberculosis etc. Pulmonary function test was initially done and the results showed abnormality. She was advised

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to take chest X-ray and the result showed increased diffuse reticulonodular shadow. On series of investigation, her liver and renal functions were normal and tested for rheumatoid and the relative factors were negative. She was admitted in the hospital for further investigations.

Diagnosis and Treatment

To identify the type of disease, various diagnosis procedures were carried out High Resolution Computer Tomography (HRCT) was done and the report showed honeycombing and basal fibrosis in lungs. There were many findings in HRCT: a) Honey combing b) apical, emphysema c) traction bronchiectasis (mainly in the basal and sub pleural regions) d) scarring of lung parenchyma. Lung biopsy was not done due to severe functional impairment of the patient. The range of clinical parameters of normal patient is given in Table. 1. The Initial PET results of the patient discussed in this case study are shown in Table 2.

Table 1. PET Test values for Normal patient

Parameters	May 2014
VC% Predicted	80% to 120%
FRC % predicted	75% to 120%
RV %Predicted	75% to 120%
DLCO	>60% to 120%
FVC%predicted	80% to 120%
FEV1%predicted	80% to 120%
FEV1/FVC	75% to 85%

Table 2. PFT Test Result

Parameters	May 2014	August 2015	July 2016	June 2017	Dec2018	Feb 2019
VC% Predicted	99	103	78	78	74	70
FVC% Predicted	104	107	77	79	73	69
FEV1% Predicted	97	96	79	80	77	70
FEV1/VC Predicted	69	70	63	68	55	48
DLCO-SB	99	76	54	57	42	31
DLCO-VA					54	43
Heart Rate/Min			72	70	132	145
Oxygen Saturation	99	95	88	88	89	62

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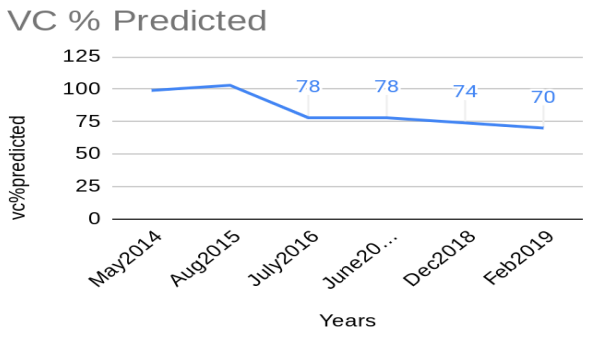
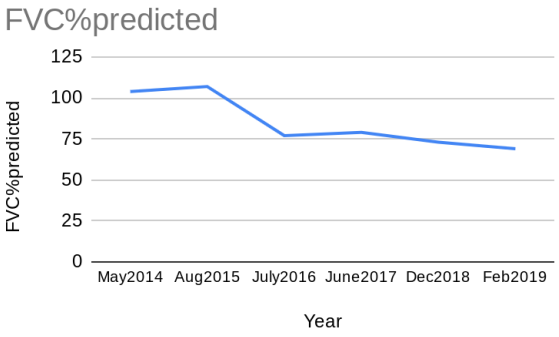


(a)



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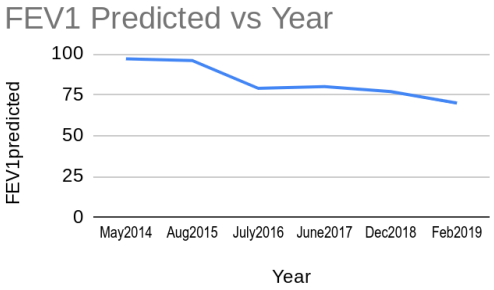

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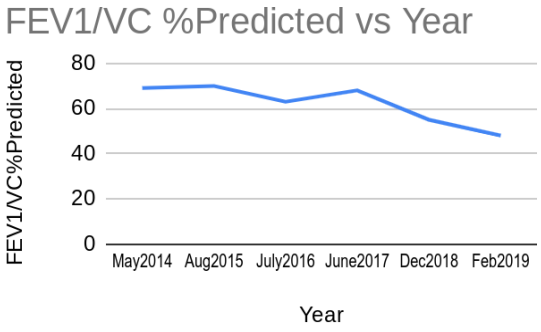

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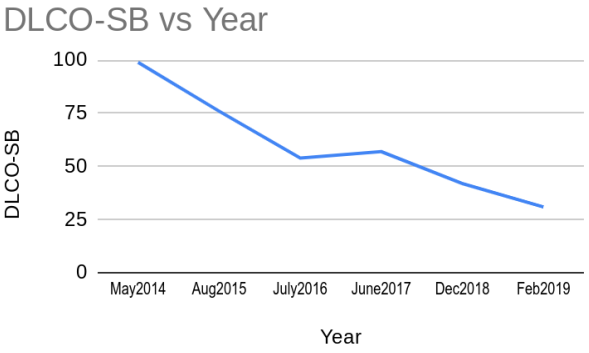

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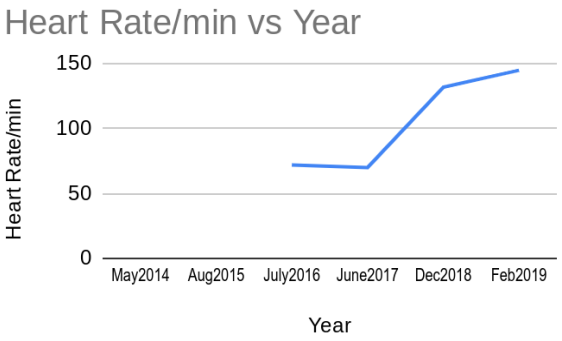

(f)

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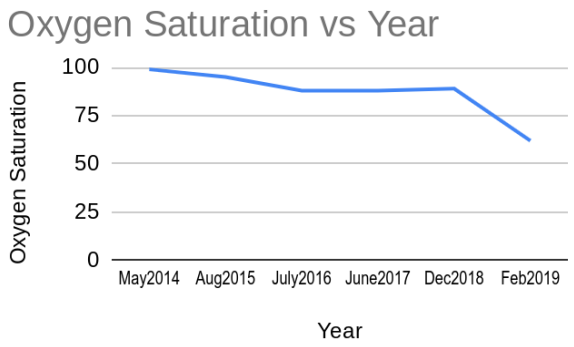

(g)

Fig. 2 pulmonary function tests from 2014 to 2019 (a)- vital capacity (b) Forced vital capacity (c) Forced Expiratory Volume (d) FEV1 (e) diffusing capacity of the lung for carbon monoxide (DLCO) (f) Heart Rate/min (e) oxygen saturation (arrow indicates patient was receiving supplemental oxygen). Start: oxygen saturation at the beginning of the 6MWD; end: oxygen saturation at the end of the 6MWD.

The Woman had a continuous cough with a sputum and had a 500 m walk test. The patient was given steroid with a dosage of 20 mg and it was gradually decreased to 10 mg. because of side effects. There were no improvement in her dyspnoea on exertion had deteriorated progressively. PFTs performed continuously and the results revealed that there were no stabilisation of DLCO and FEV1, FVC and VC

Result and Discussions

Interstitial lung disease (ILD) is a very broad term for over 200 different diseases that includes variation in terms of clinical process, treatment and prognosis. Among these various pulmonary fibrosis, detecting appropriate management and protecting prognosis is very critical. Idiopathic pulmonary fibrosis (IPF) is the most common of the idiopathic interstitial pneumonias and has a worse prognosis than other ILD. From the above case study, it is noted that the survival rate ranges from 3 to 5 years only even if the patient underwent various treatments and procedures. The patient was diagnosed in the year 2014 and she was under medication and follow-up treatments by pulmonologists. Even Though, her condition was worsened year by year and sudden flare up was in the month of December 2018. She passed away in 2019 march. The patient's initial PFTs taken in 2014 revealed a number of variations in the parameters including VC (99% pred), FVC (104% pred), FEV1/VC(69% pred) and DLCO (99% pred). Resting oxygen saturation was normal (99%). But on continuous assessment, we can see the fast variation in the parameters year by year. In Feb 2019 , the results were VC (70% pred), FVC (69% pred), FEV1/VC(48% pred), DLCO-SB(31% pred) and DLCO-VA(43% pred). Resting oxygen saturation was normal (60%) and the heart beat/min (145). For such patients, the only solution is to regularly monitoring their pulmonary function parameters and if required radiological examinations.

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Conclusion

ILD patients should be monitored and treated at regular intervals and if required apart from medication some radiological procedures should be carried out including physiological treatment. However, the physician identifies that the progression of disease through declined representations and an accelerated growth of disease, the intervene of treatment in the ILD. Many of them might think that early detection of rapid progress is very much important for the patients to decide the type of treatments. Our patient was a young female, non-smoker and non-contact with ILD causing parameters.

Limitations and Future Studies

ILDs caused by unknown cause's type cases can be analysed and finding alternative procedures to identify the sudden/progressive growth of the disease might help the patient's life smoother and longer.

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