

# Idiopathic Pulmonary Fibrosis with Complications at Nnamdi Azikiwe University Teaching Hospital, Nnewi, Nigeria: A Case Report.

**Anyabolu AE<sup>1</sup>, Enemuo EH<sup>1</sup>, Ele PU<sup>1</sup>, Ugoeze FC<sup>1</sup>, Ufoaro CU<sup>1</sup>, Nwagbara CT<sup>1</sup>, Ukah CO<sup>2</sup>, Onwukamuche ME<sup>2</sup>, Ndukwe CO<sup>2</sup>.**

## ABSTRACT

**Background:** Idiopathic Pulmonary Fibrosis is a progressive diffuse lung disease, characterised by interstitial fibrosis, minimal inflammation, and architectural distortion interspersed with areas of normal lung tissue. It is rarely reported in Nigeria.

**Aim:** The aim is to present a case of idiopathic pulmonary fibrosis in our centre.

**Methods:** A review of a case of a 60 year old woman who presented to our centre with two year history of progressively worsening breathlessness and unproductive cough.

**Results:** A 60 year old foodstuff trader was managed in our center with two year history of progressive breathlessness and unproductive cough. She eventually developed complications of right sided heart failure and hypoxemia. The investigations she had included spirometry, chest X-ray, pulse oximetry, high resolution chest CT scan and lung necropsy. The results are all in keeping with idiopathic pulmonary fibrosis. She died after about 8 months steroid therapy as well as antifailure regimen.

**Conclusion:** Idiopathic pulmonary fibrosis is a chronic progressive fibrosing interstitial pneumonia of unknown cause. Lung transplant is the only effective treatment as response to medical therapy is generally poor.

**Key Words:** Idiopathic Pulmonary Fibrosis, Right sided heart failure, Hypoxemia

## INTRODUCTION

Idiopathic Pulmonary Fibrosis (IPF) is a specific form of chronic progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, limited to the lungs and associated with the histopathologic and/or radiologic pattern of Usual Interstitial Pneumonia (UIP)<sup>1</sup>. It is the most common

form of idiopathic interstitial pneumonia<sup>2</sup> and the type with worst prognosis<sup>3</sup>. The prevalence of idiopathic pulmonary fibrosis in the USA is 14 per 100,000 and incidence of 6.8 per 100,000<sup>4</sup> but data is scarce on the epidemiology of IPF in Nigeria.

Development of IPF is influenced by both genetic and environmental factors. Family history of pulmonary fibrosis is strongly associated with increased risk of IPF<sup>5</sup>. Also, studies have shown increasing association with cigarette smoking<sup>6,7</sup>, environmental exposure to birds and cats<sup>8</sup>, gastro-esophageal reflux disease<sup>9,10</sup>, viral infection<sup>11</sup> and occupational exposure to metal and wood dust<sup>12</sup>.

There is currently no evidence that any drug treatment improves survival of patients with IPF<sup>13</sup>. The prognosis of IPF is poor, with an estimated median survival of 2-5 years from diagnosis<sup>14,15</sup>.

## CASE REPORT

A 60 year old female Nigerian of Ibo origin, who was a foodstuff trader, presented with a 2-year history of insidious onset progressive breathlessness and non productive cough. There was no history of fever, weight loss, night sweat, or contact with person having chronic cough. She had no chronic exposure to organic or inorganic dust. Though dyspnoea progressed to occur at rest, there was no orthopnea, paroxysmal nocturnal dyspnoea or leg swelling. She had no history of tobacco use.

Her physical examination findings included respiratory distress with respiratory rate of 28 cycles per minute; widespread fine end inspiratory crackles, pulse of 100 b/min and blood pressure of 140/80mmHg. Other systems were essentially normal.

Significant investigation findings included: Negative sputum smear examination for AFB; ESR of 4mm/hr; initial SPO<sub>2</sub> of 94% and negative HIV screening test. Her spirometric result, as shown in table 1, depicted a restrictive pattern. Chest X-ray result showed loss of volume with crowding of ribs on the right, compensatory hypertrophy of the left lung, patchy opacification of most lung fields with reticulonodular background (figure 1).

Figures 2 to 5 are serial axial cuts of the patient's high resolution CT of the chest from the apex to the base. It shows diffuse involvement of both lungs with more apical sparing and basal pathology. There are reticulonodular opacities, marked peripheral interstitial

<sup>1</sup>Respiratory unit, Department of Internal Medicine, Nnamdi Azikiwe University Teaching Hospital, Nnewi, Nigeria.

<sup>2</sup>Department of Histopathology, Nnamdi Azikiwe Teaching Hospital, Nnewi, Nigeria.

**Corresponding Author:** Dr. Ugoeze Francis C. Respiratory Unit, Dept. of Medicine, Nnamdi Azikiwe University Teaching Hospital, Nnewi.  
E-mail: ociso@yahoo.com.

thickening, cystic airway disease (mainly in the posterior segment) volume loss (more on the right) and regions of ground glass opacification characteristic of idiopathic pulmonary fibrosis.

She was commenced on tabs prednisolone 40mg daily (after meal), caps omeprazole 20mg daily, oral antibiotics when there was evidence of chest infection and supplemental oxygen when SPO<sub>2</sub> is < 92%. She was followed up fortnightly to monthly. She however progressively deteriorated despite treatment as she ultimately developed grade 2 digital clubbing as well as bilateral leg edema, raised jugular venous pulsation of 7cm, tender hepatomegaly and ascites suggestive of predominant right sided heart failure. She was managed in ICU as SPO<sub>2</sub> dropped to 63% but later died

after 3days in ICU.

Lung necropsy result (figure 6 and figure 7) showed patchy interstitial fibrosis of varying intensity (ie some fibroblastic foci and other areas of dense collagenization) with interspersed areas of relatively normal lung parenchyma. The fibrotic areas show moderate inflammation consisting mostly of lymphocytes and a few plasma cells, neutrophils and eosinophils. It also showed alveolar destruction with cystic changes (honeycomb fibrosis) with smooth muscle hyperplasia and pulmonary oedema. All these are in keeping with usual interstitial pneumonia.

**Table 1: Spirometry result.**

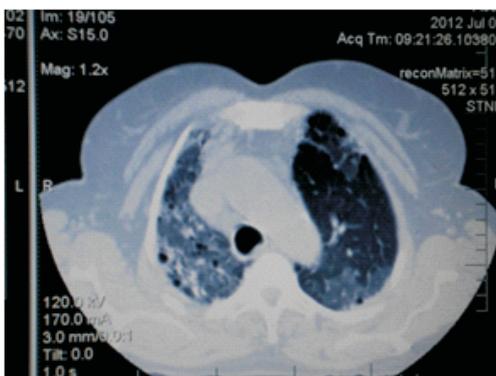
	PRED	Measured	%Measured
FVC	2.12	1.21	57
FEV <sub>1</sub>	1.78	0.84	47
FEV <sub>1</sub> /FVC	78.8	69.4	88
PEF	5.63	2.88	51
FEF <sub>2575</sub>	2.98	0.29	10



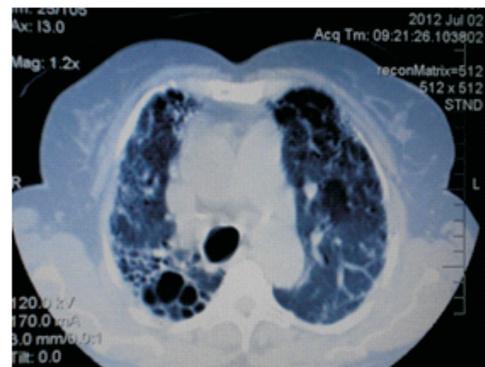
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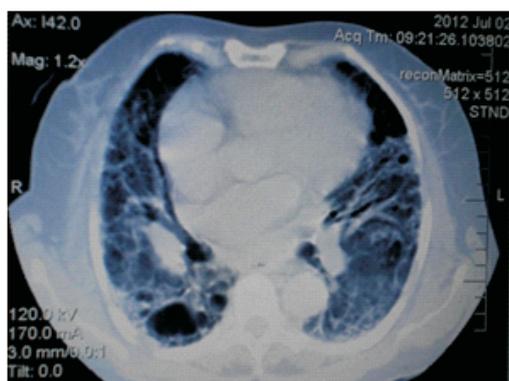
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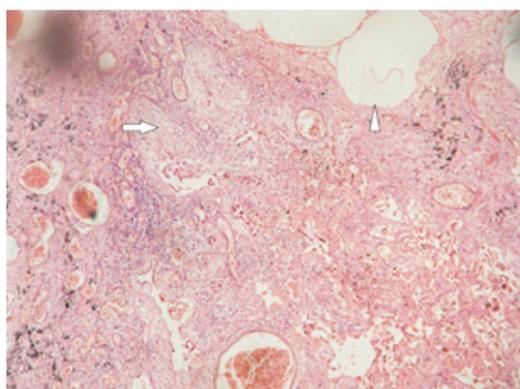
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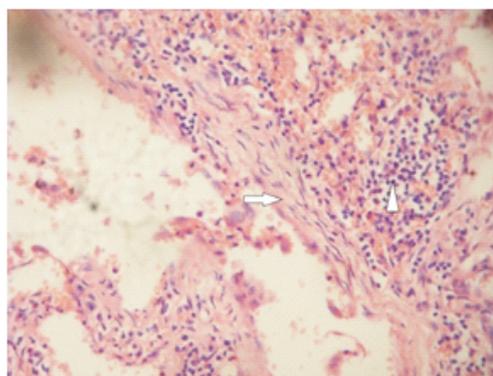
D



E



G



F

### Legends to Figures:

- A: Figure 1, showing the chest X-ray of the patient.
- B: Figure 2, apical axial cut of the chest CT.
- C: Figure 3, axial cut showing ground glass opacity (L) with normal lung tissue.
- D: Figure 4, axial cut showing subpleural honeycombing.
- E: Figure 5, axial cut at the lung bases.
- F: Figure 6, photomicrograph (H&E), arrow represents fibroblastic foci, arrowhead represents foci of chronic inflammatory cells.
- G: Figure 7, photomicrograph (H&E), arrow represents areas of fibrosis, arrowhead represents honeycombing.

## DISCUSSION

Idiopathic pulmonary fibrosis is commoner in older adults with the highest incidence in persons more than 50 years. In the United States of America, the age-adjusted incidence rate by both narrow-case and broad case finding criteria in a community based study was higher in men than in women with highest incidence among those aged 70-79 years<sup>16</sup>.

Data are scarce on prevalence of idiopathic pulmonary fibrosis in Nigeria. This report represents one of the few cases of IPF reported in Nigeria and perhaps the first case confirmed by tissue diagnosis from South-Eastern Nigeria.

A surgical lung biopsy is required for definitive diagnosis of IPF. Limitation to extensive investigations in Nigerian patients with IPF (including lung biopsy) could partly account for the paucity of work on IPF in Nigeria. Also, the frequency of autopsy in tertiary centers is low due to many factors including sociocultural beliefs.

However, a diagnosis of IPF can be considered likely in the absence of a surgical lung biopsy specimen when certain major (4) and minor criteria (3) are met<sup>14</sup>. This patient was extensively investigated with all findings in keeping with the diagnosis of IPF. Tissue diagnosis in

this patient was possible probably because of her demise. The diagnosis of IPF in few published work done in Africa are based on high resolution CT scan<sup>8,17</sup>. There is no effective therapy for IPF. Corticosteroids alone were the mainstay for the treatment of IPF but the response to treatment is generally poor<sup>18</sup>. Combined corticosteroid and cyclophosphamide therapy has no impact on survival in patients with IPF<sup>19</sup>. Lung transplant is the only treatment with an impact on survival, but it concerns only a minority of patients and must be performed early in the disease process<sup>20</sup>. Piferidone, a novel antifibrotic and anti-inflammatory drug, reduces deterioration in the lung function and therefore represents an appropriate treatment option for patient with IPF<sup>21</sup>.

Our patient was treated with steroid for about a year before she died. Her deterioration despite steroid therapy appears to corroborate the reported poor treatment response to steroids and poor prognosis of idiopathic pulmonary fibrosis.

## CONCLUSION

Idiopathic pulmonary fibrosis is a chronic progressive fibrosing interstitial pneumonia of unknown cause commoner in older adults. It is rarely reported in Nigeria partly limitations due to extensive investigations required to establish the diagnosis. Lung transplant is

the only effective treatment as response to medical therapy is generally poor.

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