Rare disease

A rare case of cor pulmonale secondary to idiopathic pulmonary fibrosis in Nigeria

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Summary

Idiopathic pulmonary fibrosis (IPF) is a rarely reported disease in Nigeria. Cor pulmonale, one of the complications of this type of diffuse parenchymal lung disease is even rarer. The authors present a Nigerian patient with IPF with a classical high-resolution CT features, managed in our centre together with associated problems.

BACKGROUND

Idiopathic pulmonary fibrosis (IPF) is a rarely reported disease in Nigeria. Cor pulmonale, one of the complications of this type of diffuse parenchymal lung disease (DPLD) is even rarer.

IPF is a disease of uncertain aetiology classified in one of the four subtypes of DPLD formerly known as interstitial lung disease.¹ The DPLD affects the parenchyma primarily, thus distinguishing them from diseases that affect predominantly the airways and the blood vessels in the respiratory tract.

The current hypothesis in the pathogenesis of IPF cites exposure to inciting agents (like smoke, environmental pollutants, environmental dust, viral infections, gastro-oesophageal reflux disease, chronic aspiration) in a susceptible host which leads to initial alveolar damage. Activation of alveolar epithelial cells leads to the migration, proliferation and activation of mesenchymal cells and formation of fibroblastic/myofibroblastic foci, accumulation of extracellular matrix and irreversible destruction of lung parenchyma.²

The American Thoracic Society and the European Respiratory Society emphasise the importance of clinical-radiological-histological correlation in the diagnosis of IPF. Their consensus report defines broad criteria and narrow criteria for diagnosis of IPF.³ Though, a surgical lung biopsy is fundamental in diagnosis, but a high-resolution CT (HRCT) may be diagnostic if the radiological features are classic for the disease. The classic HRCT features include coarse reticular or linear opacities in the basilar or subpleural regions of the lung, subpleural honeycomb cysts, focal or diffuse ground glass opacification, thickening of the intralobular/interlobular septae and traction bronchiectasis.³

CASE PRESENTATION

A 50-year-old male, brick layer and farmer from Nimbo, Uzo Uwani, Enugu, Nigeria was admitted on 11 December 2009 for breathlessness and cough of 4 years duration,

and bilateral leg swelling for 2 months. His clinical presentation, including physical examination findings were respiratory distress, a respiratory rate of 50 cpm, pulse of 120 bpm, cyanosis, finger clubbing, bilateral pitting pedal oedema, left parasternal heave, loud p2, grade 3 pansystolic murmur, maximal at lower left sternal edge, raised jugular venous pressure, hepatomegaly, ascities, as well as chest findings of patchy bronchial breath sounds and crepitations.

INVESTIGATIONS

Investigation findings included sputum examination for AFB-negative; chest x-ray-enlarged cardiac silhouette, opacity in the right mid and lower lung zones with obliteration of costophrenic angle. ECG showed sinus tachycardia, P pulmonale, right ventricular hypertrophy with strain pattern, right axis deviation. Echocardiography, demonstrated right ventricular dilatation with pulmonary and tricuspid regurgitation and pulmonary hypertension. Lung function test reported a restrictive pattern. HRCT showed irregular reticular opacities and regions of ground-glass opacification, honeycombing and generalised volume loss (figures 1 and 2). Figure 3 showed pulmonary artery enlargement in keeping with pulmonary artery hypertension.

TREATMENT

He was placed on intermittent oxygen 4 l/min as needed, intravenous frusemide 80 mg 12 hourly, tab spirinolactone 50 mg, intravenous ceftriaxone, subcutaneous enoxaparin 40 mg daily, tab vitamin C 200 mg thrice daily, tab prednisolone 20 mg daily.

OUTCOME AND FOLLOW-UP

Patient was discharged after modest clinical improvement – oedema resolved, milder cough and breathlessness.

The major constraint in managing this patient was finances.

Figure 1 Honeycombing.

DISCUSSION

This is perhaps the first case report of IPF and cor pulmonale secondary to IPF from Nigeria.

Worldwide, the prevalence of IPF is estimated to be 20 cases per 100 000 persons for males and 13 cases per 100 000 persons for females.⁴

Prevalence in USA, among residents aged 50 years or older ranges from 27.9 cases per 100 000 persons (narrow-case criteria) to 63 cases per 100 000 persons (broad-case criteria).⁵ Cor Pulmonale account for 10–30% of all hospital admissions for heart failure in USA and DPLD is a significant cause of the pulmonary disease.⁶

Data on cor pulmonale from Africa is sparse. Soweto Heart of Africa study reports, 27% of the subjects including 13% of black participants had right heart failure.⁷ Pneumoconiosis, sequelae of pulmonary tuberculosis, chronic pulmonary obstructive disease, infectious diseases such as schistosomiasis and AIDS, peripheral venous disease, sarcoidosis and chronic pulmonary thromboembolism

are common causes of pulmonary hypertension in adults in sub-Saharan Africa.^{8–10}

There are reports of cor pulmonale from Nigeria but IPF was not listed as a cause.¹¹ Causes of cor pulmonale in Nigeria included chronic obstructive airway disease, pulmonary tuberculosis, chronic suppurative lung disease, connective tissue disease and sickle cell disease, pulmonary atherosclerosis, kyphoscoliotic heart disease, pulmonary fibrosis, schistosomiasis and primary pulmonary hypertension.¹¹

Other reports on cor pulmonale from Ghana and Kenya did not mention IPF as a cause of heart failure.^{12 13}

IPF has a poor prognosis; it has a mean survival of 2–5 years from the time of diagnosis.¹⁴ In an average, mortality is 64.3 deaths per million in men and 58.4 deaths per million in women.¹⁵

The most common causes of death in patients with IPF include acute exacerbations of IPF, acute coronary syndromes, congestive heart failure, lung cancer, infectious causes and venous thromboembolic disease.¹⁴

Figure 2 Irregular reticular opacities and ground glass opacification.

Figure 3 Enlarged main pulmonary artery.

Anti-inflammatory agents, antifibrotic agents, immune modulators are available as treatment options for IPF but lung transplantation is the only proven beneficial modality of management.

Treatment for cor pulmonale include – oxygen therapy, diuretics, vasodilators, digitalis, theophylline and anti-coagulation.

Learning points

- The diagnostic and management hurdles surrounding IPF may have made it an under-reported disease in Nigeria.
- Studies are also required to unravel possible relationships between environmental toxins and the cardiopulmonary systems.

Competing interests None.

Patient consent Obtained.

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