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Pneumonie Interstitielle Non Spécifique chez une Femme Nigériane de 28 Ans : Défis du Diagnostic et de la Prise en Charge dans un Contexte de Ressources Limitées

[1]*B. A. Ajayi, [1]H. O. Iheonye, [2]A. A. Akor, [3]B. I. Ododo

ABSTRACT

BACKGROUND: Nonspecific interstitial pneumonia is a rare clinical condition and usually precedes the full manifestation of connective tissue disorders, drug-induced interstitial lung disease or chronic hypersensitivity pneumonitis.

OBJECTIVE: This article reports a twenty-eight (28)-year-old Nigerian female with nonspecific interstitial pneumonia (NSIP) complicated by Cor pulmonale. The unusual characteristics of our index case favors NSIP rather than idiopathic pulmonary fibrosis (age of 28 years, female, chest high-resolution computerized tomographic features and response to glucocorticoids).

RESULTS: High-resolution computerized tomography scan of the chest showed honeycombing in the apical lobes and a mixture of reticular changes, ground-glass appearance and fibrotic changes in basal lobes. Echocardiography revealed features of right chamber enlargement with pulmonary hypertension. Due to financial constraint, serological tests for CTDs and lung biopsy could not be done.

CONCLUSION: Being a rare condition, a high index of suspicion with full investigative workup to make early diagnosis and effect prompt treatment is important. **WAJM 2022; 39(12): 1316–1318.**

Keywords: Interstitial pneumonia, Interstitial lung disease, Connective tissue disease, Cor pulmonale, Resource constraint, Young female.

RÉSUMÉ

CONTEXTE: La pneumonie interstitielle non spécifique est un état clinique rare qui précède habituellement la manifestation complète de troubles du tissu conjonctif, de pneumopathie interstitielle d'origine médicamenteuse ou de pneumonie d'hypersensibilité chronique.

OBJECTIF: Cet article présente le cas d'une Nigériane de vingt-huit (28) ans atteinte d'une pneumonie interstitielle non spécifique (PINS) compliquée d'un cor pulmonaire. Les caractéristiques inhabituelles de notre cas d'index favorisent la PINS plutôt que la fibrose pulmonaire idiopathique (âge de 28 ans, femme, caractéristiques de la tomographie informatisée à haute résolution du thorax et réponse aux glucocorticoïdes).

RÉSULTATS: La tomographie informatisée à haute résolution du thorax a montré des alvéoles dans les lobes apicaux et un mélange de changements réticulaires, d'aspect en verre dépoli et de changements fibrotiques dans les lobes basaux. L'échocardiographie a révélé des caractéristiques d'élargissement de la chambre droite avec hypertension pulmonaire. En raison de contraintes financières, les tests sérologiques pour les CTD et la biopsie pulmonaire n'ont pas pu être effectués.

CONCLUSION: Comme il s'agit d'une maladie rare, il est important d'avoir un indice de suspicion élevé et de procéder à un bilan d'investigation complet afin de poser un diagnostic précoce et d'appliquer un traitement rapide. **WAJM 2022; 39(12): 1316–1318.**

Mots clés: Pneumonie interstitielle, pneumopathie interstitielle, maladie du tissu conjonctif, cor pulmonaire, contraintes de ressources, jeune femme.

[1]**Department of Internal Medicine, Federal Medical Centre Lokoja, Kogi State, Nigeria.**
[2]**Department of Medicine, University of Abuja, Federal Capital Territory, Abuja, Nigeria.**
[3]**Department of Radiology, Federal Medical Centre Lokoja, Kogi State, Nigeria.**
***Correspondence:* Dr. Bukola Anthony Ajayi,** Department of Internal Medicine, Federal Medical Centre Lokoja, Kogi State, Nigeria. P.M.B 1001. Phone number: +234 816 845 9687 Email: tonyboymed@gmail.com. Postal code: 260101.

Abbreviations: CTD, Connective Tissue Disorder;**ESR,** Erythrocyte Sedimentation Rate; **HRCT,** High Resolution Computed Tomography; **ILD,** Interstitial Lung Disease; **IPF,**,Idiopathic Pulmonary Fibrosis; **NSIP,** Nonspecific Interstitial Pneumonia.

INTRODUCTION

Interstitial lung disease (ILD) is a diverse group of diffuse parenchymal disorders characterized by varying degrees of inflammation and fibrosis. Over 200 different disorders are under the umbrella of ILD.[1]

The unusual characteristics of our patient favors nonspecific interstitial pneumonia (NSIP) rather than idiopathic pulmonary fibrosis (IPF). To the best of our knowledge, this is the first documented case of ILD in North-central part of Nigeria and the youngest patient with NSIP in Nigeria. High resolution chest computerized tomogram scan plays a central role in making diagnosis particularly when the findings are classical without need for lung biopsy to differentiate NSIP from IPF.[2, 3]

CASE REPORT

A twenty-eight (28)-year-old woman presented at the emergency room with 13 months history of progressively worsening cough. It was initially dry and later became productive of scanty whitish sputum, occasionally stained with blood and at times yellowish. The cough was not posture related, not associated with evening pyrexia, wheeze or chest tightness. There was an accompanying history of intermittent low-grade fever and chest pain. She also noticed progressive exertional dyspnea to dyspnea at rest, palpitation and progressive weight loss.

About a month before presentation, she developed progressive bilateral leg swelling and abdominal pain. She had poor sleep at night due to persistent cough and dyspnea.

There was a history of oliguria and dizziness, no fainting or irrational talk. A review of the other systems did not yield any additional information. She had no history of exposure to fumes, gases, irradiation or biomass fuels/fumes. She does not keep pets or birds. No anterior neck swelling. She is not hypertensive, diabetic or asthmatic. No known drug allergy. The menstrual period had stopped six months prior to presentation and not on any method of contraception. She does not smoke cigarettes or drink alcohol. She had presented earlier at various hospitals where she tested negative to Mycobacterium tuberculosis (gene expert and sputum AAFB) and was said to have normal chest X-ray. She took different cough expectorants and antibiotics without any improvement. She visited various traditional homes and took different herbal preparations.

Examination revealed a young woman, conscious, chronically ill-looking, in respiratory distress, not pale, not febrile (37°C), centrally cyanosed, SPO_2 in room air was 66%, grade IV digital clubbing, bilateral pitting pedal edema to the thighs with sacral edema, no peripheral lymphadenopathy, no asterixis.

Pulse rate was 120 beats/minute, small volume and regular. Blood pressure was 90/60mmHg and jugular venous pulse was elevated. Precordium was hyperactive with displaced apex beat and presence of right ventricular heave. The first, second and the third heart sounds were heard with palpable pulmonic component of second heart sound, a pansystolic murmur grade III/VI was heard at the fourth left parasternal area.

Respiratory rate was 30/minute; the trachea was central. Chest expansion was equal bilaterally; tactile fremitus and vocal resonance were equal bilaterally. Breath sound was vesicular with bi-basal inspiratory crackles.

The abdomen was grossly distended with anterior abdominal wall edema. No obvious distension of superficial abdominal veins. There was tender hepatomegaly with pulsatile liver. Ascites was demonstrable by fluid thrill. Nervous system examination was essentially normal. Erythrocyte sedimentation rate (ESR) was 60mm/hr on admission and 28mm/hr in the third week after discharge. Human immunodeficiency virus screening was negative. Complete blood count, urinalysis, serum electrolytes, urea and creatinine were essentially normal. There was unconjugated hyper-bilirubinemia and elevated serum alkaline phosphatase (424IU/L), while other liver enzymes were within normal limit. There was sinus tachycardia, right axis deviation and right bundle branch block on electrocardiography.

Pre-treatment chest x-ray posterior-anterior view (Figure 1a) showed cardiomegaly, with bilateral fibrocystic and streaky changes suggestive of honey-comb pattern. The costophrenic sulci showed hazy appearance. Echocardiogram showed moderate tricuspid regurgitation (dense and broad continuous wave doppler spectra), hugely dilated right chambers with right ventricle almost obliterating the left ventricle. There was mild pericardial effusion.

High-resolution computed tomography scan (HRCT) of the chest showed widespread reticular opacities in both lung fields with cystic changes and traction bronchiectasis more in the upper lung fields (Figure 2). The upper and lower lung lobes (bilateral) had a ground glass appearance and widespread thickening of the interlobar septa (Figure 3). The right middle lung lobe was preserved with no lesion noted within it. There was mild pleura effusion.

She was managed for Cor pulmonale from interstitial lung disease (ILD) likely nonspecific interstitial pneumonia (NSIP) with digoxin 0.125mg daily, spironolactone 25mg daily and torsemide 50mg daily. Non-sedating mucolytics and dihydrocodeine minimally reduced the severity of the cough. Due to moderate pulmonary hypertension, oral sildenafil 25mg daily was prescribed for three (3) weeks. This was later discontinued after complete resolution of peripheral edema and the cardiac silhouette had returned to normal on repeat chest radiograph (Fig. 1b). Oral prednisolone and hydroxychloroquine were commenced on strong suspicion of connective tissue disease (CTD).

Intermittent low dose oxygen at 2–3L/min was administered at an average of 10 hours daily as oxygen saturation range was 80–86% in room air. Rivaroxaban 10mg daily was commenced as prophylaxis against coagulation disorder associated with prolonged immobilization and ILD.

OUTCOME/RESULTS

She was discharged after 4 weeks on admission when peripheral edema had resolved completely. Oxygen saturation in room air improved to 90–93% with minimal need for oxygen supplementation and she was able to walk to the bathroom without support. She was seen

twice in the clinic after discharge at 2 weeks interval and was able to trek 500 meters without being unduly dyspneic. However, the dry cough persisted. Repeat chest X-ray showed remarkable improvement, but repeat echocardiogram could not be done due to financial constraint.

DISCUSSION

Few cases of ILD have been reported among Nigerians ≥ 50 years[4,5] and none below the age of 30 years. Pesonen *et al* reported atypical IPF in a 39-year-old African.[6] Several factors could contribute to the low incidence among Nigerians/Africans. Racial/ethnic differences in presentations have been suggested [-6] and challenges in making an accurate diagnosis exist because hi-tech investigative modalities such as chest HRCT scan are not readily available. In addition, tissue histology has been difficult because of scarcity of material resources for the procedure and associated complications unless during post mortem. Widespread poverty also limits the extent of evaluation that can be done even when the facilities are available because of exorbitant cost for out-of-pocket policy instead of health insurance. The latter factor played a major role in our index case.

Nonspecific interstitial pneumonia (NSIP) has two histological subtypes: fibrotic and cellular subtypes.[1] The common features on imaging are relatively symmetric and bilateral ground-glass appearance, associated fine reticulations and traction bronchiectasis. Presence of immediate subpleural sparring is specific for NSIP, however honeycombing is unusual.[1–3] NSIP is usually seen in the younger age group and is typically associated with an underlying clinical disorder, particularly CTD, unlike idiopathic pulmonary fibrosis which is typically seen in older individuals > 60years.[1,2]

Histology plays a central role in making an accurate diagnosis as overlapping features between the variants of NSIP and IPF has been reported.[1,3] In our index case, clinical parameters (< 30years of age, female gender and elevated ESR) and the presence of predominant ground glass opacities in all lobes except the right middle lobe, favor a diagnosis of NSIP rather than IPF.[1]

In addition, she made significant clinical and radiological improvement following commencement of steroid. This also supports the possibility of NSIP which has a more favorable outcome following use of steroid with or without azathioprine and mycophenolate mofetil.[1]

From the extensive history and physical examination, we could not identify any clue to any CTD. Financial constraint prevented further evaluation for systemic lupus erythematosus, mixed CTD and dermatomyositis/polymyositis in a young female with significantly elevated ESR. It has been documented that ILD could manifest years ahead of typical features of a particular CTD.[1,2] Elevated serum bilirubin and alkaline phosphatase could be from chronic hepatic congestion. Lung biopsy was considered but could not be carried out because of recurrent hypoxemia and the patient did not consent to the procedure.

This report highlights the existence of ILD in Nigerians, particularly among young patients with probably NSIP in Africa. It is the first documented case in the North-central part of Nigeria and possibly the youngest case of NSIP in Africa to the best of our knowledge. It also mentioned the challenges encountered in the evaluation of such a patient in a limited resource setting.

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Conflict of Interest

None.

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None.

Previous publication/presentation

None.

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