Yellow nail syndrome – report of a rare disorder

[1]Peter Ogie Ikuabe, [2]Ikenna Desmond Ebuenyi, [3]Olugbenro Osinowo, [1]Nathan Ebimobowei Onini

Abstract

Background: The Yellow Nail Syndrome (YNS) is a rare disorder of unknown cause characterized by the trail of yellow and thickened nails, lymphoedema and respiratory manifestation. We have no record of any case report of yellow nail syndrome in our setting.
Methods: We reviewed the records of an 80 year old woman who presented with two weeks history of exacerbation in her three months history of cough, leg swelling, shortness of breath and left sided chest pain.
Results: She was found to have thickened and yellow nails, lymphoedema and pleural effusion. The pleural fluid had an acellular smear with Lactate dehydrogenase(LDH) of 2,214U/L, glucose was 11.9mmol/L and protein of 59g/L and culture yielded growth of *streptococcus pneumoniae.*
Conclusion: This case report highlights the need for a high index of suspicion in making such a diagnosis.

Key words: Yellow nail syndrome, pleural effusion, leg swelling.

Highland Med Res J 2017;17(1):65-67

Introduction

The yellow nail syndrome (YNS) is a rare disorder of unknown cause characterized by the triad of yellow and thickened nails, lympheodema and respiratory manifestation [1, 2, 3]. It was first described by Samman and White [3] in 1964, and approximately 150 cases have been reported in the literature, consisting of small case series or isolated case reports. The pathogenesis, clinical presentations and natural history of this disorder remain, for the most part, obscure. Available data suggest acquired lymphatic dysfunction to be the predominant mechanism underlying the clinical manifestation of YNS [1].

Although Samman and White[3] are usually credited for introducing the term 'Yellow Nail Syndrome' and providing the original description of the disorder, the first reported cases can probably be attributed to Hiller[1] who in 1927 described two patients with abnormal yellow nails and peripheral oedema. In a review article on nail disorders published in 1962. Samman and Strickland[4] presented 41 patients with various nail abnormalities, four of whom also had evidence of lymphoedema, a condition thought to result from impaired peripheral circulation. Samman and White[3] published the first case series of YNS in 1964. Thirteen cases were presented, all having in common the presence of thickened yellow and slow – growing (<0.25mm/week) nails, in association with lymphoedema in 10 of 13 patients. Lymphangiographic data were presented for four of these 13 patients and were abnormal in all, revealing diffusely hypoplastic lymphatic vessels. Impaired lymphatic drainage was offered as the explanation for the diffuse peripheral oedema observed in these patients, but whether it could also explain the nail findings was unclear.

Emerson[1] in 1966 reported the association of YNS with pleural effusion, a frequent manifestation of the syndrome. Runyon, in 1979, suggested that a trail of symptoms including, yellow nails, lymphoedema and pleural effusion was the most characteristic presentation of YNS. The observation that some of these findings may resolve over time led Hiller et al[2] to suggest that the presence at any given time of two of these three manifestation was sufficient to established the diagnosis of YNS.

Case Report.

NB is an 80 year old female who presented with two weeks history of cough, leg swelling, shortness of breath and left sided chest pain. She gave a history of background hypertensive heart disease, type II diabetes mellitus and knee osteoarthritis. She did not smoke but cooked with firewood. She did not take alcohol. She was on glibenclamide, metformin and lisinopril.

Her temperature was 36.1^{0}C, pulse rate was 100/minute, respiratory rate was 30 cycles per minute and blood pressure was 170/110mmHg. She had

[1]Department of Internal Medicine, Niger Delta University Teaching Hospital, Okolobiri, Bayelsa Nigeria.[2]Athena Institute, Faculty of Earth and Life Sciences, Vrije Universiteit, Amsterdam, The Netherlands [3]Department of surgery, Niger Delta University Teaching Hospital, Okolobiri, Bayelsa Nigeria.

All correspondences to:
Ikenna Ebuenyi
E-mail: desiykee@yahoo.com

bilateral pitting pedal oedema and bilateral pleural effusion which was more marked on the left. Examination of the nails showed thickened yellow nails. First and second heart sounds were heard with a loud second heart sound in the aortic area. The abdomen and central nervous system were normal.

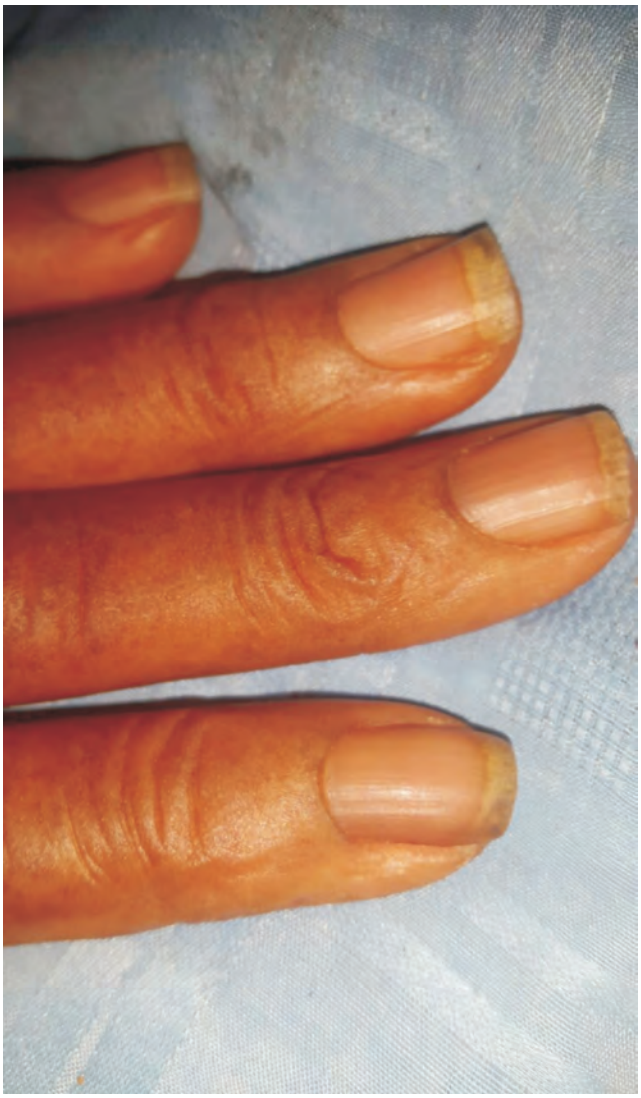

Figure 1a: Left fingers showing thickened yellow nails

Her haemoglobin was 10g/dL with a total white cell count of 3.1 x 10^9/L. Her erythrocyte sedimentation rate (ESR) was 150mm/hr. Her retroviral test, hepatitis B and C test,Veneral Disease Research Laboratory (VDRL) test were negative. The pleural fluid gave an acellular smear with Lactate dehydrogenase (LDH) of 2,214U/L, glucose was 11.9mmol/L and protein of 59g/L – Plain chest radiograph showed bilateral pleural effusion which was more on the left. Sputum was negative for acid fast bacilli but showed pus cells and culture yielded moderate growth of *streptococcus pneumoniae* after 24 hours of aerobic incubation. Her urinalysis showed glycosuria of

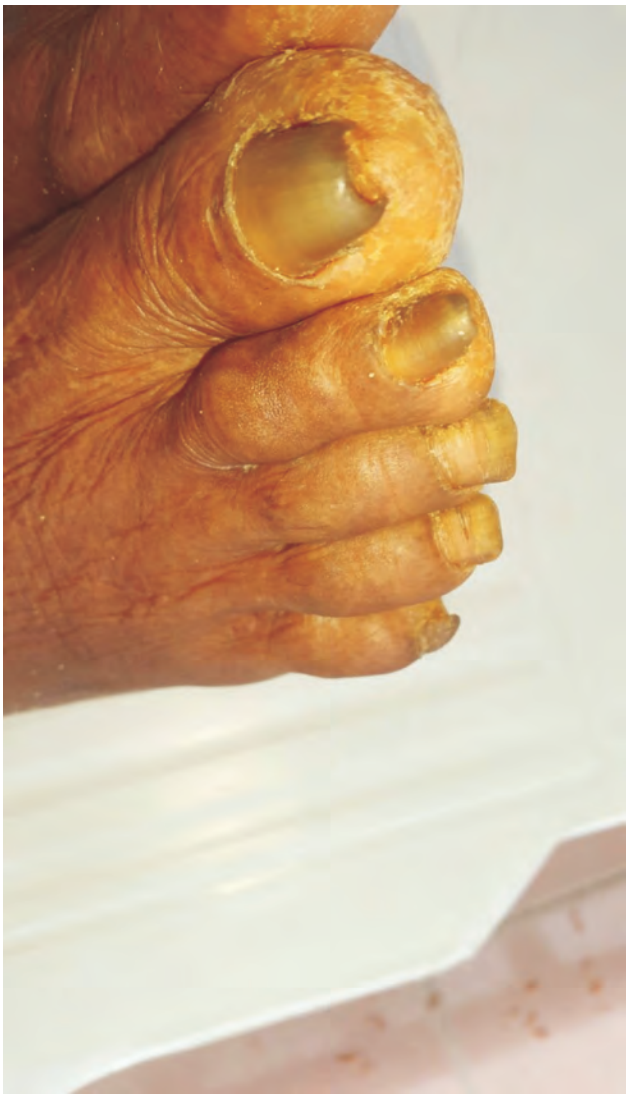

Figure 1b: Right toes showing thickened yellow nails

+++ and proteinuria of +. Her biochemistry results showed sodium of 139mmol/L, potassium 3.5mmol/L, chloride 98mmol/L, bicarbonate 24mmol/L, urea 3.8mmol/L, creatinine 111umol/L, ASAT 8U/L, ALAT 10U/L, total protein 65g/L, albumin 41g/L. Her electrocardiograph showed evidence of left atrial enlargement echocardiography was in keeping with hypertensive heart disease. She had a closed chest thoracotomy tube drainage. The diabetes was controlled on 10iu of soluble insulin 8 hourly and her blood pressure was controlled on lisinopril at 20mg daily. She had pleurodesis done and was discharged home.

Discussion

The initial lymphangiographic characteristics of four of the 13 patients described by Samman and White[3] in 1964 constitute the cornerstone of this syndrome. Nordkild et al[5] in 1986 reviewed the lymphangiographic data available in the medical literature and noted that

anatomic abnormalities of the lymphatic ducts were noted in the majority of cases (15 of 18 patients) and included lymphatic hypoplasia, dilations and extensive collateral lymphatic network. These anatomic abnormalities, however are not universally described in cases of YNS and can be difficult to reconcile with the frequent improvement of peripheral edema over time, a phenomenon rarely encountered in other forms of lymphoedema. Our patient had peripheral fluid collection in form of pedal oedema when she was not in heart failure nor hypoproteinaemic. Functional lymphatic abnormalities have been suggested as a more likely explanation and have been documented in some cases of YNS through lymphoscintigraphy. Bull et al [6] reported 17 cases of YNS of whom nine had lower extremity swelling and two had features of lymphoedema.

Runyon et al [7] analyzed pleural fluid turnover using a protein-bound dye and demonstrated decreased lymphatic flow to be responsible for the development of pleural effusion and appeared to support the notion of functional lymphatic abnormality in YNS. The biochemical feature of pleural effusions in YNS, characterized by a high protein content, as in our patients, but transudative by all other criteria, would be consistent with this explanation. Histologically, Solal-Celigny et al [8] reported findings in a patient with YNS after a pleurectomy was performed for a recurrent pleural effusion in 1983. As in our patient, the pleura showed thickening and chronic inflammation with dilated lymphatic capillaries, similar to previous reports.

In a recent case series, pleural biopsy specimens were available for seven of 41 patients [9]. Although chronic pleural inflammation was again noted, lymphatic vessels appeared grossly normal. Functional lymphatic disorder, rather than structural disease, is currently favoured as the shared pathogenic mechanism for the development of pleural effusions and lympheodema.

The nail manifestations are more difficult to explain solely on the basis of lymphatic dysfunction. Ectatic endothelium-lined vessels, possibly lymphatic in nature, have been described in the nail bed of patients with YNS [10]. The nail manifestation of YNS appear to vary over time and have occasionally been reported to evolve in parallel with the respiratory manifestations of the syndrome[8,9] . Some workers have suggested that oxidation of liquids in the nail plate may lead to accumulation of lipofuscin, a pigment responsible for the characteristic discoloration of the nails [10].

The mechanism of recurrent upper and lower airway infections frequently encountered in this syndrome remains unclear. Impaired lymphatic drainage at the microcirculation level may delay clearance of bacteria and promote microbial proliferation, ultimately leading to the well recognized complications of bronchiectasis and chronic sinusitis observed in this disease [10]. Our patient, on presentation, produced sputum from which *streptococcus pneumoniae* was cultured.

The management of pleural effusion is usually tailored to the size of the effusions, symptoms and clinical context [9,10]. Therapeutic thoracenteses may suffice in controlling symptomatic pleural effusion with pleurodesis as our patient had. Yellow Nail syndrome is a rare disorder and in our environment, no previous case has been reported. We found it good enough reason to report this one.

One limitation of this report is the differential diagnosis of post-pneumonic pleural effusion but the long-standing history makes this unlikely. Her serum albumin and total serum protein were within normal limits making hypoproteinaemia as cause of the effusion to be unlikely

Author Contributions

POI ,OO and NEO were involved the patient management. POI conceptualized the paper and conducted the literature search with IDE. POI,IDE, OO and NEO were involved in writing and editing the draft. All the authors read and approved the final manuscript before submission.

Acknowledgement

The authors are grateful to patient for giving consent and to the doctors and health personnel in departments of Internal medicine and surgery at Niger Delta university Teaching Hospital Bayelsa.

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