

Case Report

Yellow Nail Syndrome - A Diagnostic Dilemma

Vidushi Jain¹, Ashish K Jaiswal², R N Singh³

¹Assistant Professor, Department of Skin & VD, Mulayam Singh Yadav Medical College & Hospital, Hapur, U.P.;

²Assistant Professor, TB & Chest, Mulayam Singh Yadav Medical College & Hospital, Hapur, U.P.;

³Associate Professor, Department of Microbiology, Mulayam Singh Yadav Medical College & Hospital, Hapur, U.P.

ABSTRACT:

A case of a yellow nail syndrome is described in a fifty year old male patient who presented with classical triad of this syndrome i.e., deformed yellow nail, lymphedema and chronic bilateral recurrent pleural effusion. After extensive workout along with thorascopic pleurodesis, none of the associated factors was elucidated and therapeutic trial of Anti Tuberculous therapy was given. After nine month of therapy complete resolution of Pleural effusion was seen.

Key words: yellow nail, pleural effusion, lymphedema.

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Corresponding Author: Dr. Vidushi Jain, Assistant Professor, Department of Skin & VD, Mulayam Singh Yadav Medical College & Hospital, Hapur, U.P., India

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INTRODUCTION

Yellow nail syndrome (YNS) is a rare disorder, characterized by rhinosinusitis, pleural effusions, bronchiectasis, lymphedema, and dystrophic yellow nails.¹ The classic triad, described by Emerson, of lymphedema, slow-growing yellow nails, and pleural effusion is seen in only one third of patients.² Individual manifestations of the syndrome can appear at different times, and clinical onset varies from birth to late adult life.³ Yellow nail syndrome has been associated with autoimmune disorders, such as thyroiditis, systemic lupus erythematosus, and rheumatoid arthritis. There is also isolated case reports of Yellow nail syndrome associated with malignancies in cancer of the breast, larynx, lung, endometrium, gall bladder, metastatic sarcoma, metastatic melanoma, Hodgkin's disease, and mycosis fungoides. It has also been described in tuberculosis, AIDS, and other immunodeficiency states, and with the use of certain drugs.^{4,5} Pulmonary Tuberculosis has not been associated with the yellow. There is only one case where it proved to be a putative factor.⁶

This is an infrequently reported clinical entity with only few cases reported from India. The present report describes a case of Yellow nail syndrome that had

all the classical features of this syndrome and which also resolves by course of Chemotherapy.

CASE REPORT

A chronic smoker of 56 year with history of variable breathlessness and swelling of both legs and feet for eight months presented with increased breathlessness for 10 days. He had mild morning cough with small quantity clear viscid phlegm but had no chest pain, orthopnea, haemoptysis, fever or any other systemic complaints. Clinical examination revealed bilateral below knee lower limb pitting edema (Figure 1), and thick, yellow discolored nails of fingers and toes (Figure 2), and reduced breath sound on right lower chest. The nails are yellow and excessively curved both longitudinally and laterally (Figure 3). Cardiac auscultation and JVP were normal. There was no organomegaly or enlarged lymph node. Since eight months, he had to undergo repeated pleural fluid aspirations, three times from right and twice from left. At consultation he had moderate right and mild left pleural effusion (Figure 4). 1000ml of yellow clear fluid was aspirated. Biochemical report of all aspirated fluid was similarly lymphocytic exudate. His hemogram, sugar profile, HIV screening, LFTs, HbsAg, ECG, echocardiography and abdominal sonography were

normal. His sputum and pleural fluid for AFB were also negative by ZN and PCR. Filarial antigen test was negative. CT chest showed bilateral pleural effusion with normal pulmonary paranchyma and chest skeletal (Figure 4). Pleural biopsy revealed fibro-collagenous tissue with few chronic inflammatory cells. Nails scrapings for fungus on KOH mount were also negative. He was diagnosed to have YNS and anti-tubercular therapy was instituted for ninth months. This was followed by partial recovery in nails and complete resolution of pleural effusion.



Figure 1: showing below knee lower limb pitting edema



Figure 2: Showing thick, yellow discolored nails of fingers and toes



Figure 3: Showing yellow and excessively curved nails

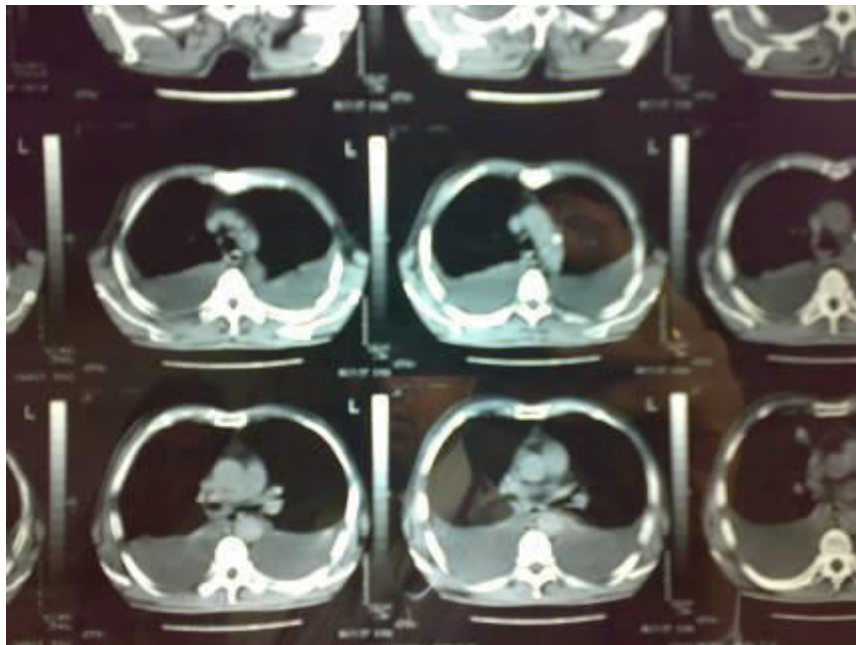


Figure 4: Showing bilateral pleural effusion with normal pulmonary paranchyma and chest skeletal

DISCUSSION

Pleural effusion, although a common clinical entity in clinical practice, poses a diagnostic challenge when of chronic or recurrent nature. Yellow nail syndrome is rare cause of such presentation with associated yellow nails and lymphedema. Some physician restricts the diagnosis to patients who have the complete triad, whereas others accept any two characteristics.

The basic abnormality of this syndrome appears to be hypoplasia of lymphatic vessels which is responsible for lymphedema, nail changes and pleural effusion. In overview of 97 patients, lymphedema was first identified in neonates or in patients as old as 65 years with the mean age was 40 years. 89% of patients had yellow nails, 80% had lymphedema of varying severity and 36% had pleural effusion.⁷

Airway manifestations include rhinosinusitis and bronchiectasis. Yellow nails result from slow growth, possibly secondary to defective lymphatic drainage. The nails become dystrophic with longitudinal or transverse ridging and loss of lunula and cuticle and they are typically yellowish in colour and thickened.⁸ The lymphedema is often mild, usually affects the lower extremities and can be demonstrated by peripheral lymphangiography.^{9,10} The pleural effusion is bilateral in about 50% and varying in size from small to massive. Pleural effusions appear to be a later manifestation of the syndrome secondary to inadequate drainage by overstressed hypoplastic lymphatics rather than increased fluid production.^{9,10} Pleural fluid is usually clear exudate with lymphocyte predominance.^{11, 12} Once a pleural effusion has developed with this syndrome, it tends to persist and recur rapidly after

Thoracentesis.¹² If the effusion are persistent and the underlying lung can be re-expanded, chemical pleurodesis or pleural abrasion can be done to prevent re-accumulation of fluid. If the lungs fail to re-expand the drainage of the pleural space can be achieved by pleuro-peritoneal shunts.¹³ The cause of bronchiectasis is unclear, but again, dysfunctional lymphatics are thought to play an important role with compromised drainage of secretions and local immune function.

The diagnosis is made when a patient has a chronic pleural effusion in conjunction with yellow nails or lymphedema. The case presented here showed all the features of classical triad of Yellow nail syndrome. In our country diagnosis may be difficult or missed at times as patient may not present with all features of this syndrome simultaneously or when present with each aspect of the syndrome in the different departments. Further more, the nail changes in Indian patients may not be easily appreciable because of colour of skin. Lymphedema and chronic pleural effusion can easily be attributed to filariasis and tuberculosis respectively as both these diseases is endemic in our country. Increasing awareness of this syndrome and close scrutiny of the nails in patients having idiopathic or recurrent pleural effusion and/or lymphedema of legs will avoid diagnostic delay and other necessary treatment modalities.

In the literature there was only one case review by shiu Pang⁶ where Tuberculosis is the causative factor for Yellow nail Syndrome. In the above case the complete recovery of effusion after tuberculous treatment strongly suggest that Tuberculosis is the underlying cause of Yellow nail syndrome.

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