Case Report

Yellow Nail Syndrome

*Junior resident, **Senior Consultant Pulmonologist, Department of Pulmonary Medicine, Chest Hospital Calicut

Correspondence: Dr. Sanjeev Shivashankaran, Junior resident Chest Hospital, Pavamani Road Calicut - 4 drsanjeev1210@gmail.com

Abstract:
Yellow nail syndrome (YNS) is a rare condition defined by the presence of yellow nails associated with lymphedema and/or chronic respiratory manifestations. Here we report a case of yellow nail syndrome who presented with pleuritic chest pain and pedal edema.

Key words: Yellow nail syndrome, lymphedema

Introduction:
Yellow nail syndrome (YNS) is a rare disorder first described in 1964 by Samman and White, consisting of yellow nails with lymphedema and/or chronic respiratory manifestations. The respiratory manifestations are diverse and include pleural effusion, bronchiectasis, rhino sinusitis, chronic cough, or recurrent lung infections. Here we present a case report of 58 year old female patient who came with pleuritic chest pain and pedal edema and was diagnosed to have yellow nail syndrome.

Case report:
A 58 year old female diabetic patient presented to our hospital with complaints of left sided chest pain, loss of appetite, fatigue and bilateral leg swelling. On examination patient had bilateral pedal edema and signs of left sided pleural effusion. Chest radiography (Fig 1) confirmed the presence of left sided pleural effusion. Thoracocentesis was performed and pleural fluid study showed exudative

Fig (1):
Chest x ray showing left sided pleural effusion
effusion which was lymphocyte predominant with low ADA. Gram stain and AFB stain was negative for organisms. There was no evidence of malignant cells. Her Echo cardiography, Anti nuclear antibody (ANA) Profile and Thyroid Profile were normal. As we could not proceed with thoracoscopy due to patient’s unwillingness we made the patient undergo CT Thorax which showed left sided pleural effusion with no obvious parenchymal abnormality (Fig2). So we started her on empirical anti tuberculosis treatment.

![CT showing left sided pleural effusion](image1)

*Fig (2) : CT showing left sided pleural effusion*

On her next follow-up her symptoms did not improve and her edema was persisting. So we repeated a chest X ray (Fig 3) and CT Thorax (Fig4) which showed bilateral pleural effusion. So we made her undergo thorough general and systemic examination. General examination showed thick, curved yellowish nails of fingers (Fig 5,6) and toes (Fig 7) with non pitting edema of both lower limbs and left upper limb (Fig 5,7). A KOH preparation of nail scrapings was negative for fungus. Based on the findings we made a final diagnosis of yellow nail syndrome. Patient was started on diuretics and vitamin E. After one month, patient became symptomatically better and is currently under follow up.

**Discussion:**

The initial description of Yellow nail syndrome in 1964 by Samman and White included 13 subjects with dystrophic slow growing nails and lymphedema. On the basis of lymphangiographic studies obtained in four of these patients, they hypothesized that the
(8) Guillain-Barre’ syndrome
(9) Xanthogranulomatous pyelonephritis
(10) Tuberculosis
(11) Myocardial infarction
(12) Nephrotic syndrome
(13) Exudative enteropathy
(14) Hypoalbuminemia
(15) Drugs (thiol compound therapy)

Pathology:

Lymphatic dysfunction is thought to be involved in the pathogenesis of YNS. Nail fold capillaroscopy studies in YNS have occasionally shown dilated and tortuous capillary loops, a finding suggestive of microangiopathy as the cause for the nail changes. Impaired lymphatic drainage in the bronchi and bronchioles has been suggested as a cause for recurrent infections and development of bronchiectasis. Runyon et al. analyzed pleural fluid turn over 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 but transudative by all other criteria, would be consistent with this explanation. Few case reports of familial YNS have been described; an autosomal dominant pattern of transmission is sometimes mentioned in the literature, in spite of scarce evidence to support YNS as an inheritable disorder. Most cases are sporadic with clinical manifestations occurring relatively late. Very few pediatric cases have been reported.

Clinical features

1. Abnormal nails: Yellow discoloration
   Slow growth (<0.25mm/week)
   Abnormal thickening
   Transverse ridges
   Excessive curvature
   Onycholysis

2. Lymphedema: Lower extremities, upper
extremities or face. Hypoplastic or dilated lymphatic ducts or both on lymphangiography.

Delayed lymphatic drainage on lymphoscintigraphy.

3. Respiratory manifestations:
   Cough or shortness of breath.
   Pleural effusions (chylous or non chylous)
   Bronchiectasis
   Chronic sinusitis

Diagnostic criteria:

The diagnosis of YNS is essentially a clinical one and based on the presence of characteristic findings including abnormal nails, lymphedema and respiratory manifestations that may include pleural effusion, bronchiectasis and sinusitis. The early recognition that some of the manifestations of YNS are inconsistent and variable over time has led to the general consensus that two of the three manifestations of YNS may be sufficient to strongly suggest the diagnosis in the absence of another plausible explanation.

Treatment:

1. Respiratory manifestations of bronchiectasis can be controlled with a combination of postural drainage and other bronchopulmonary hygiene measures, in combination with the judicious use of antimicrobial therapy. Optimal control of bronchiectasis with bronchopulmonary hygiene measures (postural drainage, chest physiotherapy, use of flutter valve, inhaled bronchodilator if indicated), influenza and pneumococcal immunizations, and prompt treatment of complicating respiratory infections are advisable.

2. The management of pleural effusion occurring in patients with YNS should be tailored to the size, persistence, and clinical effects of the effusion. Those patients with recurrent symptomatic accumulation of pleural effusion may require pleurodesis or other surgical maneuvers.

3. Although the efficacy of vitamin E remains unclear, it seems reasonable to offer this option to patients who are troubled by their nail manifestations.

4. Lymphedema can be managed in most patients with a regimen that consists of gradient pressure garments, exercise, bandage wraps, and manual lymph edema drainage or external pneumatic compressions as needed. Overall, our results suggest that the manifestations of YNS can be managed with currently available means in most patients and progressive respiratory insufficiency appears to be uncommon.

Conclusion

The diagnosis of yellow nail syndrome 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. Furthermore, the nail changes in Indian patients may not be easily appreciable because of color of skin. Lymph edema and chronic pleural effusion can easily be attributed to filariasis and tuberculosis respectively as both these diseases are endemic in our country.

References:


11 Hoque SR, Mansour S, Mortimer PS. Yellow nail syndrome: not a genetic disorder?

