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Case Report

Yellow Nail Syndrome - Report of a Classical Case from Eastern India

Arijit Nag[1]. Sumantro Mondal[2], Avinandan Banerjee[1], Sandipan Halder[1], Debanjali Sinha[1], Atanu Chakraborty[1], Tony Ete[1]

[1]M.B.B.S, post graduate trainee, Department of General Medicine, Institute of Post Graduate Medical Education And Research, Kolkata, India.
[2]M.B.B.S, D.M.R.D, post graduate trainee, Department of General Medicine, Institute of Post Graduate Medical Education And Research, Kolkata, India.

Corresponding Author: Sumantro Mondal

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ABSTRACT

Yellow nail syndrome is a rare disease characterized by classical triad of primary lymphedema, recurrent pleural effusion and yellowish discoloration of nails. There is very few reported case of this rare disease with all three classical features from India. We report a case of fifty year old male who presented with classical triad of deformed yellow nail, primary lymphedema with recurrent bilateral pleural effusion and finally diagnosed as yellow nail syndrome.

Key words: Yellow nail syndrome, classical triad, elderly male.

INTRODUCTION

Yellow nail syndrome (YNS) is characterized by the triad of deformed yellow nail, primary lymphedema and respiratory disorder including pleural effusion.[1] The exact etiology is unknown. Dysfunctional hypoplastic lymphatics may have some pathogenic role.[2] Samman and White first described the disease in 1964 with features of primary lymphedema and yellowish discoloration of nails.[3] Pleural effusion is added later as a feature of this syndrome. There are few reported cases of YNS from our country and all three classical features may not be seen in a single patient. This classical case report is associated with all three cardinal features of yellow nail syndrome.

CASE REPORT

A 50 year male admitted with recurrent cough and breathlessness for the past six years associated with gradually progressive swelling of both lower limbs for last ten years. He also noted yellowish discoloration of finger and toe nails even before the onset of lower limb swelling. He was non-diabetic and normotensive without any past history of tuberculosis. Earlier he was hospitalized twice for cough with breathlessness and bilateral lower limb swelling. Chest X rays during his first and second admission showed unilateral (right) & bilateral (Right>Left) mild pleural effusion respectively. Mantoux test, sputum for Acid fast bacilli (AFB) and sputum culture were negative at that time. Respiratory symptoms responded well with

conservative medications but lower limb swelling never subsided. A recent Chest X ray, done ten days prior admission showed bilateral (Right>Left) mild pleural effusion. At that time patient did not respond to conservative medication.

General examination revealed, Mild pallor, tachypnea (Respiratory rate-22/min, regular) and bilateral nonpitting pedal edema .The nails of all four limbs were yellowish, thickened and curved (Figure1a&b). Examination of respiratory system revealed, dull percussion note and diminished breath sound at bilateral mid and lower lung fields. Blood examination showed Hemoglobin-10.4gm/dl. Liver function test was normal apart from hypoproteinemia (4.8gm/dl) and hypoalbuminemia (2.3 gm/dl). Routine examination of Stool and d-xylose test was normal. Mantoux test, sputum for AFB and culture was negative, Blood tests for HIV, HBsAg, anti HCV & filarial antigen were negative. Chest X-ray showed bilateral moderate amount of pleural effusion (Figure 2). X ray of paranasal sinuses, ECG & echocardiogram reveals no abnormality .Abdominal ultrasound scan was normal. Doppler ultrasound of lower limb showed bilateral normal venous flow. Nail scraping for fungal growth was negative.

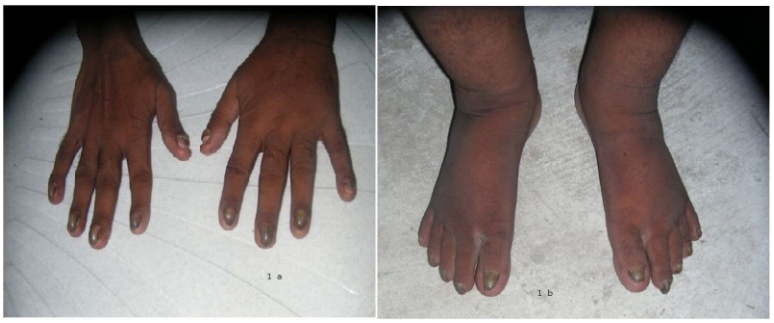

Figure 1a &b:
Photograph of hands (1a) and lower limbs(1b) of the patient showing typical curved,brownish-yellow discoloured nails,with bipedal edema.

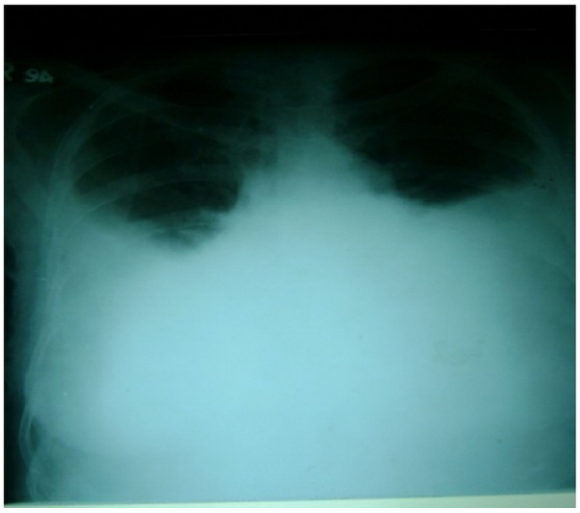

Figure 2: Chest X-Ray PA view of the patient showing Bilateral Pleural Effusion.

Diagnostic pleurocentesis from right side revealed straw colored fluid. Pleural fluid study showed protein-3.3gm/dl, sugar-98mg/dl, cell count-14/mm3(Lymphocyte-68%, polymorphonuclear-leucocytes-32%).Pleural fluid protein to serum protein ratio was more than 0.5 and pleural fluid lactate dehydrogenase (LDH 287 IU/L) to serum LDH(306 IU/L) ratio more than 0.6, compatible with diagnosis of exudative pleural effusion. Adenosine deaminase, Cholesterol, triglyceride & Amylase level of the pleural fluid were normal. Pleural fluid was negative for AFB, mycobacterial growth, pyogenic organisms or fungi. Pleural biopsy also revealed normal findings. A Chest CT Scan after thoraocentesis revealed bilateral moderate

pleural effusion with passive collapse of both lower lobes. Bronchoscopy was unremarkable. Bronchial aspirates were negative for bacterial, mycobacterial growth and malignant cells.

Repeated aspiration of pleural fluid failed to improve patient's symptoms, Tetracycline pleurodesis was done after closed tube thoracostomy on right side. During discharge pleural effusion was markedly reduced & dyspnoea was improved. In this case recurrent pleural effusion, bilateral non-pitting pedal edema and yellow nail make the diagnosis of yellow nail syndrome after exclusion of other possible causes.

DISCUSSION

Yellow nail syndrome is usually present with primary lymphedema, recurrent pleural effusion & yellowish discoloration of nail. Two of these symptoms are required for the diagnosis, the complete triad is only observed in about one-third of patients.[4] The three cardinal features of YNS may be manifested at varying times.[5] The basic abnormality of this syndrome is thought to be due to lymphatic dysfunction which is responsible for lymphedema, pleural effusion & nail changes.[6] Lymphangiogram may demonstrate lymphatic stasis in lower extremities, pelvis and abdomen.[5] Since the original description by Samman and White many association of YNS have been described. This syndrome may be associated with congenital hypoplastic lymphatics, autoimmune diseases, various malignancies and some chronic infections.[6] Association of YNS with protein losing enteropathy has also been reported.[7] This may be due to lymphatic leakage of protein and increased capillary permeability of the walls of villi. This can explain hypoproteinemia in our patient without any evidence of hepatic or renal abnormality.

Nail changes in YNS are usually manifested by, yellowish green discolouration, Onycholysis, thickened & excessively curved nail but the pathogenesis of nail changes may not be secondary to lymphatic abnormalities.[8,9] Nail changes in our patient had similar features. Pleural effusion YNS is usually exudative, lymphocytic predominant & bilateral in majority of cases. Effusion may persist for prolonged period or recur repeatedly after thoracocentesis.[10] The character of pleural effusion in our patient was consistent with these features. In case of recurrent pleural effusion chemical pleurodosis or Pleuro-peritoneal shunt is considered.

CONCLUSION

In our case, the diagnosis of Yellow nail syndrome was quite obvious, as the patient presented with all three cardinal features of the syndrome and other possible causes of pleural effusion and bipedal lymphedema were excluded. The diagnosis is sometimes difficult, especially when all three cardinal features are not present simultaneously in one patient. In India, where both Tuberculosis and Filariasis are very much prevalent, one needs to be very careful to exclude these two common diseases before making such rare diagnosis. In patient with recurrent pleural effusion and/or primary lymphedema of unknown etiology, careful examination of nails to detect characteristic changes can help a clinician to clinch the diagnosis of YNS. This case is being reported not only for its rarity but it also reestablishes the immense value of thorough general examination for correct diagnosis of a disease.

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