

Yellow Nail Syndrome- A Case Report

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Summary:

Yellow nail syndrome (YNS) is a rare disorder characterized by chronic/recurrent rhinosinusitis, pleural effusion, bronchiectasis, lymphedema and dystrophic yellow nails^{1,2}. The classic triad of lymphedema, slow growing yellow nails, and pleural effusion, described by Emerson, is seen in only one third of patients^{3,4}.

Case report

Nazir Ahmed, 55 year old male freedom fighter, a heavy smoker for >30 years, nondiabetic, normotensive, hailing from Pirozpur, presented with one of many episodes of cough and exertional respiratory distress persisting for last 2 months. Most of the attacks (over last 17 years, few attacks/year) are initiated with a flu-like illness, followed by progressive copious expectoration and breathlessness and occasional episodes were associated with wheezing. He never had chest pain, palpitation, haemoptysis or required hospitalization for these episodes. However, almost each episode was treated with antibiotics with or without bronchodilators. He related his symptoms beginning with nail changes (Fig:1) about 17 years back, progressively making the nails thick, rough, curved, shortened and brittle. However, he never had pain, features of

Here we report a case of yellow nail syndrome presented with cough, worsening symptom of breathlessness, leg edema and nail changes, waxing and waning for about 17 years.

Key word: Yellow nail, lymphedema, pleural effusion, maxillary sinusitis.

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inflammation, itching in & around the nails, or other cutaneous manifestations anywhere in the body.



Fig.-1: Nail changes in Yellow Nail Syndrome

He had developed swelling of legs beginning (Fig:2) about 17 yrs back, declining to some extent at night and with diuretics, but never disappeared completely over the period. He never had urinary symptoms, H/O



Fig.-2: Lymphoedema in Yellow Nail Syndrome

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jaundice and history suggestive of recurrent attacks of lymphangitis / thrombophlebitis or inguinal swelling. He was repeated evaluated for oedema at home and in the neighbouring country and was negative for any specific disease and received multiple courses (at least three) of full course antifilarial treatment. He also received repeated courses of anti-fungal treatment for the nail changes.

On examination at hospital, patient was mildly dyspnoeic, afebrile, pulse 72 beats/min, BP 120/70 mm of Hg, moderate non-pitting leg oedema, neck vein not visible, normal general skin. There was clubbing, and the nails had broken cuticle, thick subungual tissue, onycholysis, longitudinally ridged and had a yellow tint to its colour. Chest examination revealed vesicular breath sound with prolonged expiration, few scattered crepitations and polyphonic rhonchi, and evidence of left sided pleural effusion. Abdominal examination was unremarkable with absence of ascitis and organomegaly. Other systemic examination findings were normal.

On Investigation, the haemoglobin was 12.6 gm%, ESR-30 mm in 1st hr, total count of WBC- 6000 / cu mm and the differential count was Neutrophil-60%, Eosinophil-02%, Lymphocytes-36%. The Fasting blood sugar was 85 mg/dl & blood sugar 2 hr after breakfast was 90 mg/dl. The serum total protein was 6.13 gm/dl and serum Albumin was 3.16 gm/dl. Tuberculin test was negative, 3 sample Sputum for AFB was negative, blood test for microfilaria and Immuno-chromatographic test for filarial was negative. The renal, hepatic and cardiac function tests were normal. The thyroid function was normal (TSH - 2.25 mIU/ ml). Urinary examination revealed no protein and microscopic examination was normal. Chest X-ray cheat PA view revealed ill defined faint opacities (Fig:3) distributed bilaterally suggestive of inflammatory lung lesions with small left pleural effusion. X-ray of the paranasal sinuses were suggestive of bilateral maxillary sinusitis. Audiometric evaluation revealed mild to moderate conductive type hearing loss in right ear and mild conductive type hearing loss in left ear. Colour doppler flow study of lower limb circulation revealed no evidence of venous hypertension. Nail scrapping for fungus was negative.

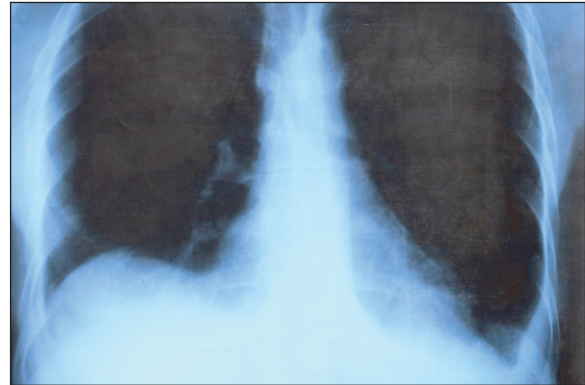


Fig.-3: *Pleural effusion in Yellow Nail Syndrome*

Ultrasonographic evaluation of abdomen revealed normal findings except for bilateral pleural effusion (left more than right). Diagnostic Pleural fluid analysis revealed deep straw coloured exudative effusion (protein 6.9 gm/dl), raised triglyceride (29 mg/dl) and polymorph pleocytosis (plenty per high power field), Few RBC was also found. Two repeat examinations of pleural fluid from both sides revealed similar findings.

Discussion

The whole team of more than 20 trainee doctors and the consultants were puzzled to find a single diagnosis for the presented clinical syndrome. He was thought to have Bronchiectasis, Allergic Rhinitis and a component of bronchial hyper-responsiveness to explain his respiratory symptoms. He was considered to have long standing Chronic Venous Insufficiency (CVI) to explain his hard pitting oedema, though post-lymphangitic lymphoedema was also on the suspicion. His nail changes was not explainable, thought to be due to fungal infection of the nails. The diagnosis was further complicated by the finding of bilateral exudative pleural effusion and at one point of time, he was a candidate for receiving a therapeutic trial of anti-TB drugs. The dermatologist's opinion regarding the nail changes were inconclusive and suggested twenty nail syndrome (non-infective) as the possible diagnosis. The trainee doctors came up with the literature search to find the Yellow Nail Syndrome to match all the components of the syndrome in our patient. Lymphangiogram could not be done to confirm lymphoedema because of non-availability of the facility in our set up or anywhere in the city. However, by exclusion we could arrive at a

diagnosis of the lymphoedema. The subsequent section will now make a brief literature review of the Yellow Nail Syndrome.

Since the original description by Samman & White³ many associated findings have been described. Yellow nails result from slow growth, possibly secondary to defective lymphatic drainage. The nails become dystrophic and dysmorphic with longitudinal or transverse ridging and loss of lunula & cuticle¹⁶. Airway manifestations include rhinosinusitis & bronchiectasis. Pleural effusion appear to be a later manifestation of the syndrome secondary to inadequate drainage by overstressed hypoplastic lymphatics rather than increased fluid production¹⁵. The cause of bronchiectasis is unclear but again the dysfunctional lymphatics are thought to play an important role with compromised drainage of secretion & local immune function¹⁴.

Yellow nail syndrome is classified as a dominantly inherited lymphedema with variable expression¹. Over 100 cases have been published, most of which have been sporadic. There have been only a few published report where a positive family history has been documented in cases of yellow nail syndrome.

The three main features of yellow nail syndrome are described in the following table².

Feature Characteristics

Nail changes (Fig:1) All nails are usually affected (though few may be spared)

Nails are slow growing or appear to have stopped growing

Nails become thicker and turn a pale yellow or greenish-yellow colour with edges slightly darker

Nails mainly remain smooth but may be cross-ridging and nail humped with loss of cuticles

Onycholysis (separation of nail from the nail bed) may affect one or more nails

Lymphoedema (Fig:2) Swelling occurs in about 80%

of patients and most frequently affects the legs

Signs of swelling usually occur after nail changes appear and may not been seen for some months later

Swelling less often affects the hands, face or genitals

Respiratory signs Pleural effusions occur in about 36% of patients

In about 30% of patients, the initial symptom is related to pleural effusions

Patients often give a history of recurrent attack of bronchitis, chronic sinusitis, and pneumonia

Individual manifestations of the syndrome can appear at different times and clinical onset varies from birth to late adult life⁵. Yellow nail syndrome has been associated with autoimmune disorders, such as thyroiditis, systemic lupus erythematosus and rheumatoid arthritis⁵. There are also isolated case reports of YNS associated with cancer of breast⁵, larynx⁶, lung⁷, endometrium⁸, gall bladder⁹, metastatic sarcoma¹⁰, metastatic melanoma³, Hodgkin's disease¹¹ and mycosis fungoidis¹². It has also been described in AIDS and other immunodeficiency states and with the use of certain drugs¹³.

Conclusion

The cause of yellow nail syndrome is unknown. However, it is seen in patients with chronic bronchiectasis or sinusitis, pleural effusions, internal malignancies, immunodeficiency syndromes, and rheumatoid arthritis. In some cases the lymphatic abnormality may be congenital (occur during development) but in most it is probably related to the other associated conditions. No specific treatment is described for the whole syndrome. Patients should receive appropriate medical treatment for their respiratory symptoms and oedema. Nail changes once established are usually permanent, although complete reversion to normal nails has been described in some of the case reports. Treatment of nails includes topical

vitamin E solution and oral itraconazole. Some studies have shown that nutritional supplementation with vitamin E appears to be effective in controlling yellow nail syndrome, for unknown reasons. Zinc supplements have also been used but it is unclear whether they are effective.

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