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## **Case Report**

# A case of pregnancy associated with the rare yellow nail syndrome: An overview and review of literature

Swati Francis<sup>1,\*</sup>, Mayadevi Kurup<sup>1</sup>, Surya Jayaram<sup>1</sup>

<sup>1</sup>Dept. of Obstetrics and Gynaecology, Aster Medcity Hospital, Kochi, Kerala, India



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## ABSTRACT

**Background:** The yellow nail syndrome (YNS) is a rare disorder characterized by the classic triad of yellow and dystrophic nails, lymphedema along with pleural effusion, resulting from malformations of the lymphatic system. The first case of yellow nail syndrome was probably reported by Heller in 1927, but Samman & White described the first series of patients who had yellow nails associated with lymphedema in 1964. <sup>1</sup> The pathogenesis of YNS still remains poorly defined. The diagnosis is established on the basis of characteristic clinical features including abnormal nails, lymphedema and any respiratory manifestations. The clinical course is usually benign, and current treatment plan aims at controlling the various clinical manifestations of this obscure disease process.

Case Presentation: Here we report an antenatal case who reported to us in her third trimester for safe confinement and delivery, already a diagnosed case of yellow nail syndrome with occasional bouts of cough on and off with no associated breathlessness. She had associated gross bilateral lower limb oedema and characteristic yellow nail changes. Physical examination revealed uterus corresponding to term gestation, relaxed and fatal heart sound good. She never had any history of associated chest pain, palpitation, haemoptysis or required hospitalization for these episodes. She gives no further association with exacerbations of any respiratory ailments.

Antenatal scan along with Doppler study revealed Breech presentation at 37-38 weeks gestation, normal Doppler, a small subserous fibroid 5x4 cms near to fundus noted. The patient was taken up for an elective caesarean section in view of the present condition of the patient and the further anticipated complications. Our case gives a glimpse of the pregnancy associated with this rare syndrome where appropriate medical treatment for their respiratory symptoms and oedema should be carried out and close monitoring of the patient is of utmost requirement in view of the anticipated complications.

**Discussion:** The yellow nail syndrome (YNS) is a rare disorder of unknown cause characterized by the triad of yellow and thickened nails, lymphedema and respiratory manifestations. The pathogenesis along with clinical presentations and natural history of this disorder remain, for the most part, obscure. Owing to the absence of large-scale studies in relation to YNS, there is no consensus on treatment strategy. Management decisions are typically based on anecdotal evidence, case reports and intuition. Treatment is predominantly supportive, but it should be further noted that successful treatment of one component of the triad often causes symptomatic improvement in the others.

**Conclusion:** The YNS remains a rare and intriguing disorder of unknown cause. Lymphatic dysfunction probably represents the pathogenic mechanism responsible for the various clinical manifestations associated with this disorder. Most clinical manifestations of YNS are generally managed symptomatically with supportive measures, and the long-term prognosis generally appears favourable.

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## 1. Introduction

The yellow nail syndrome (YNS) is a rare disorder of unknown cause characterized by the classical triad of

E-mail address: swati.francis@gmail.com (S. Francis).

<sup>\*</sup> Corresponding author.

yellow and thickened nails, lymphedema and respiratory manifestations. It was first described by Samman and White in 1964, and approximately only 150 cases have been reported in the literature so far. Hence, the pathogenesis, clinical presentations and natural history of this disorder remain, for the most part, unknown until present. Morbidity from the disease is high owing to the absence of definitive therapies for YNS and its management is primarily based on supportive and palliative therapy. In this case report, we analyse the available literature on this subject, describing clinical characteristics and its associated difficulties encountered during the antenatal and the postnatal period, associated multiple system involvements and their management. The diagnosis of YNS is essentially a clinical one and based on the presence of characteristic findings which includes abnormal nails, lymphedema and respiratory manifestations that may include pleural effusions, bronchiectasis and sinusitis along with others. 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.<sup>2</sup> Most clinical manifestations of YNS are generally manageable with supportive measures, and the long-term prognosis appears favourable.

## 2. Case Presentation

A 32-year-old lady with diagnosed case of yellow nail syndrome presented to our department for routine antenatal check-ups in her third trimester. She had previous regular antenatal check-ups which were at her hometown. Patient complained of occasional bouts of cough on and off with no associated breathlessness. There was no associated fever, body ache, bowel or bladder symptoms. She had associated bilateral lower limb gross oedema and characteristic yellow nail changes. However, patient showed no signs of exacerbation of respiratory system symptoms, the 'yellow nail' terminology only captures some of the nail changes observed in YNS. More consistent is the abnormally slow growth of the nails (<0.25mm/week), as described in the original report by Samman and White in 1964 and later confirmed by other authors.<sup>3</sup> Other nail findings include thickening, transverse ridging, excessive curvature from side to side, uneven pigmentation, diminished lunulae and onycholysis Physical examination revealed uterus corresponding to term gestation, relaxed and fetal heart sound good. She never had any history of associated chest pain, palpitation, haemoptysis or required hospitalization for these episodes.

Physical examination revealed normal pulse, BP, and temperature. Abdominal examination uterus corresponding to term gestation, relaxed and fetal heart sound good. Per speculum showed normal findings. Per vaginum examination revealed cervix soft posterior uneffaced os closed. Laboratory examination revealed marginally elevated white blood cell count with neutrophilia and C reactive protein (CRP). Antenatal scan along with Doppler study revealed Breech presentation at 37-38 weeks gestation, normal Doppler, a small subserous fibroid 5x4 cms near to fundus noted. The patient was managed by a multidisciplinary team approach including a gynaecologist, dermatologist and a general physician. Owing to the absence of large-scale studies in YNS, there is no available consensus on treatment strategy. Treatment undertaken was predominantly supportive, but it should be noted that successful treatment of one component of the triad often causes symptomatic improvement in the others.<sup>4</sup> She underwent elective caesarean section in view of the anticipated complications and the general condition and the habitus of the patient. Extracted a live term female baby of birth weight 2.8 kg. baby cried immediately after birth. Postop period uneventful with no anticipated and feared respiratory system involvement and its exacerbations. Patient was finally discharged on postop day 5 in a stable condition to review back after 1 month.

## 3. Discussion

According to study carried out by Nordkild in 1986, with a series of 97 patients who had the syndrome, the yellow nails finding was present in 99% of the cases and was the first symptom in 37% of them.<sup>5</sup> According to a study carried out by Nordkild in YNS occurs more frequently in female middle-aged patients, although it can occur from infancy to adolescence. In the case of this antenatal patient reported above, the first manifestation noted was on and off cough with expectoration, associated with bilateral lower limb lymphedema involving the feet, ankles and calves, with accentuation of the flexion folds and the characteristic yellow nails. Among the three clinical YNS characteristics (yellow nail syndrome, respiratory tract involvement, lymphedema), only two are required to diagnose YNS but it is difficult to call the entity YNS without nail abnormality.<sup>6</sup> Yellow nails and the characteristic bilateral lower limb lymphoedema are the main clinical manifestation which leads to the YNS diagnosis. However, the possible interval between the first clinical sign (lymphedema, lung manifestations) and the nail discoloration hinders affirmation of the YNS diagnosis. That yellowing represents a subset ofchromonychia, which is further defined as pathological nail discoloration, especially xanthonychia (yellow nail coloration). The nail plate becomes thickened, with an enhanced transverse curvature (overcurvature), sometimes with a notable hump associated, also associated with cross-ridging, very hard (scleronychia) and difficult-totrim nail, and cuticle disappearance.<sup>7</sup>

In case of an antenatal patient the most feared associated complication revolves around the respiratory system and its associated comorbidities the lung involvement in YNS usually noted in 56–71% of the patients, diversely affected some parts of the respiratory tract with a variety of clinical manifestations. Chronic cough is the most frequent pulmonary manifestation seen in 56% of YNS patients, with pleural effusions found in 14–46% of the patients YNS patients' pulmonary function test results are usually normal or may indicate a moderate-to-severe restrictive syndrome attributable to pleural effusion.

Lymphedema is a clinical feature of YNS, occurring in 29-80% of the reported series, and may be the first sign of the disease in about one-third of them.<sup>9</sup> Lymphedema characteristics do not differ from those of primary lymphedema. It involves the lower limbs, especially bilateral and below the knee. The increased volume of lymphoedematous limb is caused by excess lymph accumulation but also fibrosis caused by fibroblast stimulation and excess adipose tissue due to adipocyte stimulation. Acute or chronic rhinosinusitis is very common in YNS patients, estimated between 14 and 83%. 10 The maxillary sinus is the most frequently affected, followed by ethmoid, frontal and sphenoid. Yellow nail syndrome has been associated with autoimmune disorders, such as thyroiditis, systemic lupus erythematosus and rheumatoid arthritis. There are also been isolated case reports of YNS associated with cancer of breast, larynx, lung, endometrium, gall bladder, Metastatic sarcoma, metastatic melanoma, Hodgkin's disease1mphedema, pleural effusion (particularly chylothorax) or nail discoloration. Management of the respiratory symptoms in YNS patients such as sinusitis and bronchiectasis include antibiotics, bronchodilators and a good bronchial hygiene. Management of lymphedema is important in order to prevent irreversible hypertrophy of the connective tissue of the limb's patients diagnosed with idiopathic YNS may undergo a trial of medical management, beginning with oral and/or topical vitamin E, Intravenous antibiotics, and compression stockings. The largest case series suggests that life expectancy is modestly reduced when compared with that of the general population. <sup>10</sup> Progression of respiratory manifestations to respiratory failure seems very uncommon.

## 4. Conclusion

The YNS remains a rare and uncommon disorder with an unclear pathogenesis. With regards to the rare YNS, we present here with this case of antenatal mother diagnosed with the syndrome to put forward how important it is to be extremely cautious in dealing with such a case since all the associated complications, in particular involving the respiratory system have to be kept in mind and dealt with accordingly. No specific treatment is described in literature so far for this syndrome. However, it is claimed that patients should receive appropriate symptomatic medical treatment for their respiratory symptoms and oedema. Nail changes once established are usually permanent, although complete

reversion to normal nails has been noted in some of the case reports. Most clinical manifestations of YNS are generally manageable with supportive measures, and the long-term prognosis appears to be favourable.

## 5. Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

## 6. Conflict of Interest

The authors declare no competing interests.

#### 7. Authors' Contributions

All authors have read and agreed to the final version of this manuscript and have equally contributed to its content and to the management of the cases

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## **Author biography**

Swati Francis Specialist

Mayadevi Kurup Senior Consultant

Surya Jayaram Senior Specialist

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