

## Yellow nail syndrome and bronchiectasis

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### ABSTRACT

The Yellow Nail Syndrome includes slow growing, opaque yellow nails with exaggerated lateral curvature, associated with lymphoedema and chronic respiratory disorders. The nail changes may precede the lymphoedema by a number of years. Bronchiectasis may be the only chronic respiratory disorder; others include chronic bronchitis, pleural effusions and chronic sinusitis. Two illustrative cases show the combinations of some of these clinical features in two black Africans.

**Key words:** Yellow nail, bronchiectasis

### Introduction

The yellow nail syndrome (YNS) is the term coined and applied in 1964 to describe the pathological association of yellow nails and lymphoedema.<sup>1</sup> Three patients who had the combination of yellow nail syndrome and chronic recurrent pleural effusions were described two years later.<sup>2</sup> A patient with yellow nail syndrome and bilateral bronchiectasis was also subsequently reported<sup>3</sup>. In the past two decades, with increasing understanding and improved methods of investigation, the number of cases of YNS reported in the literature is increasing.<sup>4,5</sup> Within the context of current clinico-experimental knowledge, YNS is considered a clinical syndrome without specific biological parameters. The following triad of symptoms characterises it: nail abnormalities, primary lymphoedema, and pleuro-pulmonary manifestation<sup>6</sup>. It has been associated with diverse conditions including

rheumatoid arthritis,<sup>7</sup> malignancies,<sup>6,8</sup> and nephrotic syndrome<sup>9</sup>. The pathogenesis of this autosomal-dominantly inherited syndrome consists of a dysfunction of the lymphatic system<sup>10-12</sup>. In a report of the pulmonary manifestations of YNS in 12 patients, only two patients had bilateral bronchiectasis and none had the combination of bilateral bronchiectasis and sinusitis.<sup>13</sup> Two illustrative cases are presented of yellow nails associated with bilateral bronchiectasis and chronic sinusitis.

### Case reports

Case 1: A 26 year-old nulliparous female presented with 17-year history of persistent cough productive of creamy, yellowish sputum. The sputum was seldom foul smelling. She had two episodes of haemoptysis. Each episode of haemoptysis lasted about a week and the volume of blood coughed up ranged between 10 to

75mls/day. She also was worried about progressive yellow-brown discolouration and associated disfigurement of her finger- and toenails. The nails became hard, thickened, slow growing and arched. She also had onycholysis (separation of the nail from its bed) but never had evidence of peripheral oedema. She was sickly as a child during which she had severe measles infection at the age of 2 years, convulsions, tonsillitis (for which she had tonsillectomy), and chronic rhinosinusitis. Bilateral intra-nasal antrotomies did not control the symptoms of the chronic rhinosinusitis.

The features on plain chest radiographs were consistent with bilateral bronchiectasis, which were confirmed by bronchography. She had right lower lobectomy after several episodes of bronchoscopic toileting and cyclic antibiotic therapy to control the copious sputum from the poorly drained lobe. Subsequent follow-up revealed a clinically improved patient with less troublesome cough and arrest of the nail changes. She declined further surgical intervention. The histology of the excised lung was reported as bronchiectasis.

Case 2: This 54 year-old retired police officer, para 4+0, presented when she was 27 years old with chronic cough that dated to early childhood. There was an early childhood history of an almost persistent rhinorrhoea. This was diagnosed as chronic sinusitis at early adult life. She declined surgical intervention for its treatment. She was more worried about the nail changes of her hands

and feet and the bilateral ankle oedema that subsequently became prominent because of the prolonged upright posture demanded by her profession. This pedal oedema did not resolve after her second pregnancy. During the subsequent two pregnancies, she needed hospital admissions for worrisome oedema of the legs. Treatment consisted of elevation of the limbs and use of pressure stockings. Productive cough had been a common ailment she suffered since early childhood. It was preceded by measles infection. Nail changes started during her second pregnancy and became progressively worse until she started to notice her nails fall off. They seemed not to grow any more at a particular stage.

Evaluation and investigation revealed bilateral bronchiectasis, involving the lower lobes, the right middle lobe, anterior segment of the right upper lobe (Figure 1), and the segments of the lingula (Figure 2). Repeated microbiologic examination of the sputum for acid-fast bacilli was negative. After control of pulmonary secretions by bronchoscopic toileting and administration of cyclic antibiotics, she had left lower lobectomy (the most affected lobe). The histologic finding of the resected lung was consistent with bronchiectasis. She had a stormy post-operative period, and benefited from regular postural drainage and chest physiotherapy. She has been on cyclic antibiotic therapy for 27 post-operative years. The pedal oedema has improved considerably with use of pressure stockings which she wears every night. The nails have not been of concern in the past 10 years.

Figure 1: Right lung bronchiectasis



Figure 2: Bronchiectasis involving left lung



## Discussion

Yellow nail syndrome (YNS) is known as an autosomal-dominantly inherited syndrome, which consists of a dysfunction of the lymphatic system.<sup>10-14</sup> The syndrome is known to be associated with multiple organ system affectations, although three symptoms characterise it.<sup>6,15</sup> These include nail abnormalities (yellow nails), primary lymphoedema, and pleuro-pulmonary manifestations.<sup>6</sup> Currently, these symptoms are supposed to be due to impaired lymphatic drainage<sup>12</sup> consequent upon lymphatic dysfunction.<sup>15</sup>

In the development of lymphoedema in YNS, while some cases were categorised to as congenital because they developed at birth,<sup>16</sup> the majority become clinically evident only sequel to situations which may be regarded as precipitating factors these include injuries, insect bites, infections (e.g. erysipelas) or factors that cause overload of the lymphatics<sup>1</sup>. Such precipitating factors accounted for the pedal lymphoedema in one of our patients in whom lymphatic overload was the likely explanation. This seemed to be related to the onset of her profession as a police officer and the persistence of the pedal oedema of pregnancy even after delivery. The same mechanism of lymphatic overload by an infective precipitating factor may account for the pleuro-pulmonary manifestations such as pleural effusion<sup>2</sup> or recurrent respiratory tract infection.<sup>2,3,13</sup> Patients with YNS also have increased susceptibility to chest infections and recurrent cough.<sup>16-18</sup> All the 12 patients in a particular series<sup>13</sup> and both of our patients had pulmonary symptoms and recurrent cough as manifestations of their illness.

The mechanism of the pathogenesis of the bronchiectasis in YNS is less clear. The lymphatics probably play a greater role in the bronchial mechanics than had been appreciated. It has been hypothesised that abnormality in pulmonary lymphatic flow, which characterises the YNS, could result in decreased lung tissue compliance. The decreased lung compliance could account for consequent development of lower lobe pulmonary cysts associated with YNS.<sup>19</sup> A similar mechanism could be hypothesized for the

development of bronchiectasis. The repeated airway infection and ineffective lymphatic drainage around the peri-bronchial tissue, could alter the peripheral airway compliance and integrity, and lead to consequent bronchiectasis.

In a recent review of 17 patients with YNS, the majority (14 patients; 83%) suffered severe rhinosinusitis. The frequent association of rhinosinusitis with yellow nails may warrant its recognition as part of the YNS.<sup>5</sup> In a series of 12 patients with YNS who had pulmonary symptoms as manifestations of their illness, five patients had bronchiectasis and four patients had rhinosinusitis.<sup>13</sup> Rhinosinusitis occurred together with unilateral bronchiectasis in three patients. Two patients with bilateral bronchiectasis had no associated rhinosinusitis and a patient with rhinosinusitis had no associated bronchiectasis. Our report probably represents an unusual presentation in which rhinosinusitis and bilateral bronchiectasis occur together. This may well indicate delayed presentation of YNS in which bilateral bronchiectasis and rhinosinusitis represent its advanced stage.<sup>13</sup> The two patients we presented came to hospital after about two decades of respiratory symptoms and they represent a late stage in the progression of YNS.

There does not seem to be a definite order in the appearance of the characteristic symptoms of YNS<sup>1,2</sup>. More often, it may remain unrecognised in the absence of the classic triad of clinical features.<sup>4</sup> The relevance and place of high resolution computed tomography (HRCT) and standard computed tomography (CT) in the evaluation of bronchiectasis in cases of YNS has been established.<sup>20</sup> When a high index of suspicion, familiarity with the various manifestations of YNS and the features of the radiological findings are taken together, there might be an increase in the rate of diagnosis of YNS.

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