Yellow nail syndrome: Dystrophic nails, peripheral lymphedema and chronic cough

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A case involving a 41-year-old man with yellow nail syndrome (YNS) is reported. YNS is a rare disorder characterized by yellow, dystrophic nails, peripheral lymphedema and bronchiectasis with recurrent lower respiratory tract infections. YNS is often misdiagnosed because the syndrome is not well known. An interdisciplinary approach is required to recognize and collate the components of the syndrome accurately. Correct diagnosis is of utmost clinical importance because YNS can occur secondary to malignancies and autoimmune disorders. Hence, the diagnosis of YNS must prompt further investigation.

Key Words: *Bronchiectasis; High-resolution computed tomography; Tree-in-bud pattern; Yellow nail syndrome*

Le syndrome des ongles jaunes : les ongles dystrophiques, le lymphœdème périphérique et la toux chronique

Le cas d'un homme de 41 ans ayant le syndrome des ongles jaunes (SOJ) est exposé. Le SOJ est un trouble rare caractérisé par des ongles jaunes et dystrophiques, un lymphœdème périphérique et une bronchiectasie ainsi que des infections récurrentes des voies respiratoires inférieures. Le SOJ est souvent mal diagnostiqué parce qu'il est mal connu. Une démarche interdisciplinaire s'impose pour bien dépister et colliger les éléments du syndrome. Il est d'une importance clinique considérable de poser le bon diagnostic parce que le SOJ peut se produire à cause de malignités ou de troubles auto-immuns. Ainsi, le diagnostic du SOJ doit susciter une exploration plus approfondie.

Yellow nail syndrome (YNS) is a rare disorder that is characterized by the following: yellow dystrophic nails, peripheral lymphedema and bronchiectasis with recurrent lower respiratory tract infections. The disease is often misdiagnosed because the syndrome is not well known. An interdisciplinary approach is required to recognize and collate the components of the syndrome accurately. Correct diagnosis is of utmost clinical importance because YNS can occur secondary to malignancies and autoimmune disorders. Hence, the diagnosis of YNS must prompt further investigation.

CASE PRESENTATION

A 41-year-old man presented to the outpatient clinic of the University Medical Centre (Regensburg, Germany) complaining of discolouration and slow growth of his nails, leg swelling and chronic cough. He had first noticed the symptoms approximately five months before presentation. The patient's history revealed recurrent infections of the respiratory tract and pneumonia. There was no history of smoking or allergies. The nail changes had been interpreted as onychomycosis by his general practitioner and the patient had been prescribed antimycotics and biotin supplementation; however, the treatment was unsuccessful. Further evaluation of his leg swelling was performed with duplex sonography of the veins but revealed no venous abnormality.

On examination, the patient exhibited dystrophic yellowish nails and mild peripheral lymphedema. A chest x-ray revealed a consolidation in the lingula consistent with bronchopneumonia (Figure 1). High-resolution computed tomography of the chest performed at a follow-up visit revealed multifocal consolidations as well as multifocal mild cylindrical bronchiectasis with mucus impactions (Figures 2 and 3). A tree-in-bud pattern indicative of chronic bronchitis and acute infectious bronchiolitis was observed. Transbronchial biopsy was performed and showed no evidence of sarcoidosis, vasculitis or malignancy. Routine laboratory blood tests were within normal ranges. Serology for HIV, perinuclear and cytoplasmic antineutrophil cytoplasmic antibodies, antinuclear antibodies and rheumatoid factor were negative.

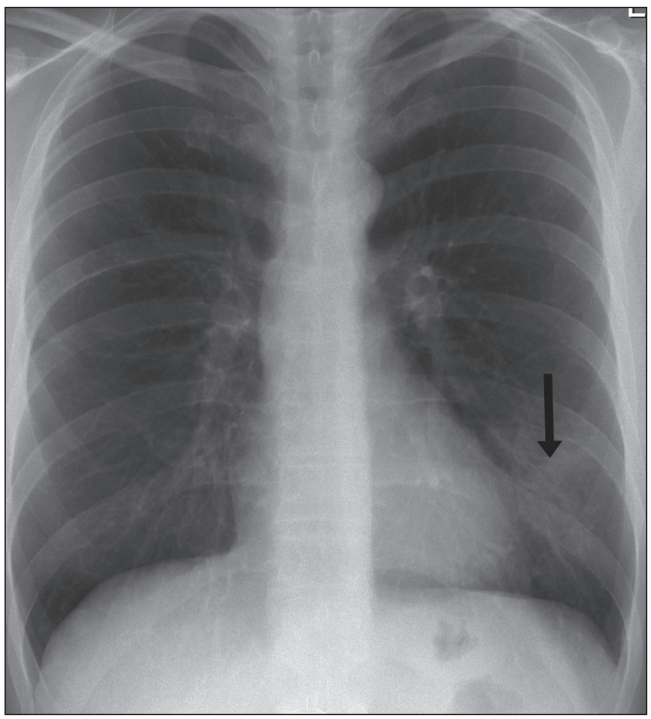

Figure 1) *Chest x-ray demonstrating a consolidation in the lingula representing pneumonia (arrow)*

DISCUSSION

Considering the triad of yellow dystrophic nails, lymphedema and bronchiectasis with recurrent lower respiratory tract infections, YNS was diagnosed. YNS is characterized by slowly growing, dystrophic,

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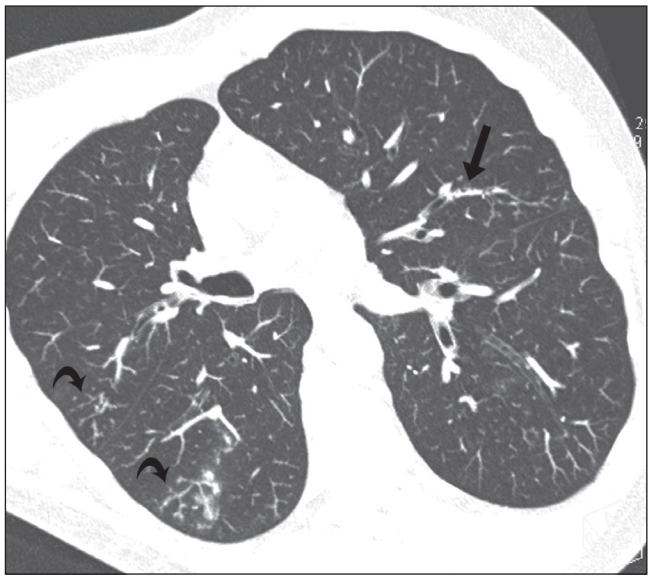

Figure 2) *The oblique maximum intensity projection of the computed tomography of the chest shows a tree-in-bud pattern indicative of infectious bronchiolitis (curved arrows) and bronchiectasis with mucus impaction indicative of chronic bronchitis (straight arrow)*

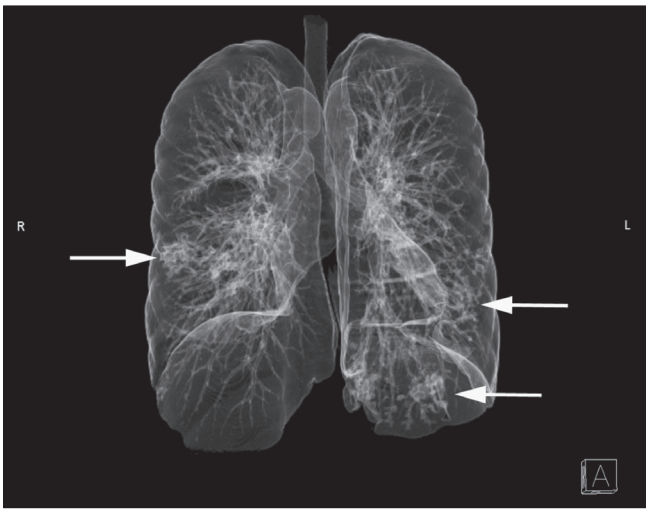

Figure 3) *Volume rendering technique of chest computed tomography demonstrating multifocal consolidations and tree-in-bud pattern (arrows)*

yellow nails, peripheral lymphedema, pleural effusions, rhinosinusitis and bronchiectasis with consecutive recurrent lower respiratory tract infections (1,2). Individual manifestations of the syndrome can appear at different times, and clinical onset varies from birth to late adult life (3). The diagnosis is made clinically and no specific treatment is known. The prognosis of YNS is generally good and supportive measures such as rotating antibiotic therapy, thoracentesis and corticosteroid therapy are often helpful in controlling the symptoms of the disease (4).

The pathogenesis of YNS remains unclear; however, anatomical or functional lymphatic drainage abnormalities have been proposed as the underlying cause (4,5). Systemic inflammatory reactions induced by recurrent respiratory tract infections may be the initiating factor for abnormal lymphatic function and clinical manifestations (4). However, YNS has been associated with autoimmune disorders (eg, thyroiditis, systemic lupus erythematosus and rheumatoid arthritis) and malignancies (eg, cancer of the breast, larynx, lung, endometrium, gall bladder, metastatic sarcoma, metastatic melanoma, Hodgkin's disease and mycosis fungoides) (4,5). YNS has also been described in patients with tuberculosis and AIDS (5).

YNS is a challenging entity that is often misdiagnosed. An interdisciplinary approach is recommended to recognize and collate the components of the syndrome accurately. The diagnosis of YNS should prompt further investigation for malignancies and other associated diseases.

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